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Paediatric Surgical Special Edition

EDITORIAL

In short order the COVID-19 pandemic brought an abrupt end to in-person congregation, discourse and learning, events which are at the very core of spreading knowledge and experience, bringing diverse communities together from across the globe.

This at a time when the provision of Global Surgery has been gaining traction around the world, care whose implementation is long overdue. Fortuitously, by virtue of high volumes of index paediatric surgical workload, carried out in academic training centres at an extremely high standard, South Africa is exceptionally well placed to contribute significantly to the Global Surgical "pandemic" in a most meaningful manner.

On account of the unpredictability around hosting in-person meetings, it was thus with some trepidation that we embarked on hosting our recent "Global Paediatric Surgical Congress". That said, the meeting proved to be a wonderful success, bringing a multitude of international delegates together to share their experiences, improving knowledge and fostering new relationships.

This Special Edition of the South African Journal of Surgery gives a broader voice to a number of the topics discussed, amongst others, vitally highlighting the wealth of knowledge and experience that the last 30 months have stolen from us. Enjoy the read!

Jerome Loveland

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PAEDIATRIC SURGERY

Coagulopathy in neonatal surgery

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Background: Neonatal patients planned for surgical intervention regularly await coagulation screening prior to surgical intervention. This study identified risk factors for coagulopathy in neonates undergoing surgical interventions to rationalise coagulation screening for patients in resource-restricted environments.
Methods: Records of neonates undergoing a surgical procedure at the study institution between June 2015 and September 2019 were reviewed. All patients younger than 44 weeks post conception were included. Age post-delivery, congenital age, reason for surgery, procedure and coagulation screen and an inflammatory marker were recorded. The patients were stratified into categories for age and surgical intervention. The rate of abnormal coagulation screens was identified for the different age and diagnoses categories and correlations were calculated between these.
Results: Two hundred and fifty-three (of 313) patients had sufficient blood results to be analysed in the study. There was no significant difference between the rate of abnormal coagulation screen according to different age categories. The necrotising enterocolitis (NEC) group had the highest rate of abnormal international normalised ratio (INR) (55%) and platelet counts (46%). A notable finding was that patients with a platelet count of > 350 are up to six times less likely to have a raised INR.
Conclusion: This study shows that the rates of abnormal coagulation screens are higher in neonates with NEC compared with other diagnoses and patients with high platelet counts (> 350) are less likely to have an abnormal INR. This is useful in resource-restricted environments to inform decision making around omitting preoperative coagulation screening in select patients to avoid unnecessary theatre delays.
Keywords: neonatal coagulopathy, neonatal surgery, necrotising enterocolitis, thrombocytosis, coagulopathy

Background

In neonates, the components of the coagulation cascade are present in different proportions to adults.[1-5] This is thought to be advantageous in this phase of life and has been described as developmental haemostasis.[1-4] During illness, the homeostasis of coagulation factors is disturbed leading to coagulopathy.[3,4] This is common in neonates with severe necrotising enterocolitis (NEC), particularly thrombocytopaenia.[6-8]

Patients who undergo surgical intervention in the neonatal phase will undergo a coagulation screen on most occasions, based on the perceived risk of bleeding in the context of sepsis or other risk factors.[9] In a resource-restricted environment, awaiting coagulation screen results can cause delays in operative intervention due to prolonged turnaround times. Delays in surgical intervention are known to contribute to poor outcomes.[10]

This study aimed to identify risk factors for coagulopathy in neonates undergoing surgical intervention and markers which could guide the decision to omit coagulation screening in selected patients.

Method

All neonates undergoing a surgical procedure between June 2015 and September 2019 were included in the study. Patients were included if they were younger than 44 weeks post conception.

The records of each patient who underwent a surgical procedure were assessed and the following were captured: age post-delivery (days); congenital age post-conception (weeks); reason for surgical intervention; procedure undertaken; coagulation screen: platelet count; international normalised ratio (INR) and partial thromboplastin time (PTT); and an inflammatory marker: C-reactive protein (CRP).

The researchers recorded coagulation screen and inflammatory marker results obtained in the 24 hours prior to theatre. The first blood results, sampled before optimisation, were recorded in cases where coagulopathy had been identified. In instances where incomplete panels of blood results were available for a patient, the available results were included and were used in selected calculations.

The patients were stratified into three categories according to age:

- *Term neonates* – less than 28 days old and greater than 38 weeks post conception at birth
- *Premature neonates* – less than 28 days old and less than 38 weeks post conception at birth
- *Premature infants* – older than 28 days but still younger than 44 weeks post conception at the time of surgery.

The reasons for surgical intervention were categorised into the following diagnoses: NEC, abdominal wall defects, congenital abnormalities and acquired disease.

Abnormal blood results were defined as INR > 1.5,[6] PTT > 80 seconds and a platelet count of < 100 x 10^9/L.[11]

The rate of abnormal coagulation screen results was identified for the different diagnoses and age categories. These were each reported as a proportion of patients who had results for the blood test within each group. The continuous, non-ranked values for each blood test (platelet count, INR, PTT and CRP) were arranged into sets and the Pearson's correlation coefficient was calculated between the respective sets. Where correlation was noted between a pair of blood results, further calculations to identify associations that could be used in clinical decision making were done. This was done by calculating the relative risk of a deranged result in one set of results when the other was above or below a set cut-off point.

Results

A total of 313 neonates underwent a surgical procedure during the 52-month period. Of these, 143 had a full set of blood results. Partial sets of results were available for 110 patients. The remaining 60 patients had one or less blood tests and were excluded.

The patient ages ranged from 0 days to 154 days post-delivery. The corrected gestational age of the oldest patient was 43 weeks and 3 days, and this patient was classified as a premature infant. The gestational age of the most premature infant at birth was 26 weeks post conception.

Term neonates represented most of the patients at 64% (n = 200). *Premature neonates* represented 24% (n = 62) of the patients and *premature infants* represented the remaining 16% (n = 51).

Table I summarises the rate of abnormal coagulation screen according to age classification. As the confidence intervals overlap, there is no significant difference between the rate of abnormal results from the coagulation screens done in the different age categories.

Table II summarises the rate of abnormal coagulation screen according to the underlying reason for surgical intervention. The NEC group had abnormal INR results in 55% of cases with results. The rate of abnormal INR in other diagnoses ranged from 19% to 24%. The platelet count was < 100 in 46% of patients with NEC, and it was < 100 in between 3% and 10% in the other diagnostic groups. The rate of abnormal PTT results was between 8% and 18% and there was no statistically significant difference between the diagnostic groups.

The Pearson correlation coefficient calculated for the different result sets ranged from r = 0.3478 to r = 0.1943. This demonstrated a weak to moderate correlation between

Table I: Coagulation screens according to age classification

	International normalised ratio				Partial thromboplastin time				Platelet count			
	≤ 1.5 (*n*)	Proportion ≤ 1.5	> 1.5 (*n*)	Proportion > 1.5	≤ 80 (*n*)	Proportion ≤ 80	> 80 (*n*)	Proportion > 80	≥ 100 (*n*)	Proportion ≥ 100	< 100 (*n*)	Proportion < 100
Premature neonate	26	0.51 CI (0.37–0.65)	25	0.49 CI (0.35–0.63)	39	0.80 CI (0.66–0.90)	10	0.20 CI (0.10–0.34)	35	0.67 CI (0.53–0.80)	17	0.33 CI (0.20–0.47)
Term neonate	109	0.72 CI (0.64–0.79)	43	0.28 CI (0.21–0.36)	127	0.88 CI (0.81–0.92)	18	0.12 CI (0.08–0.19)	148	0.87 CI (0.81–0.91)	23	0.13 CI (0.087–0.19)
Premature infant	24	0.75 CI (0.57–0.88)	8	0.25 CI (0.11–0.43)	31	1.0 CI (0.89–1.0)*	0	0.00 CI (0.00–0.11)*	31	0.82 CI (0.55–0.92)	7	0.18 CI (0.077–0.34)

CI = 95% confidence interval
Proportion calculated on patient with results (denominator adjusted for "no results" in each category)
*one-sided 97.5% confidence interval
Table I shows the abnormal coagulation screens as a proportion of all the coagulation screens done for each age group

Table II: Coagulation screens according to underlying diagnosis

	International normalised ratio				Partial thromboplastin time				Platelet count			
	≤ 1.5 (*n*)	Proportion ≤ 1.5	> 1.5 (*n*)	Proportion > 1.5	≤ 80 (*n*)	Proportion ≤ 80	> 80 (*n*)	Proportion > 80	≥ 100 (*n*)	Proportion ≥ 100	< 100 (*n*)	Proportion < 100
Necrotising enterocolitis (82)	34	0.45 CI (0.33–0.57)	42	**0.55 CI (0.43–0.67)**	60	0.82 CI (0.71–0.90)	13	0.18 CI (0.098–0.28)	40	0.54 CI (0.42–0.66)	34	**0.46 CI (0.34–0.58)**
Congenital abnormality (93)	62	0.81 CI (0.70–0.89)	15	0.19 CI (0.11–0.30)	67	0.92 CI (0.83–0.97)	6	0.082 CI (0.031–0.17)	79	0.96 CI (0.90–0.99)	3	0.037 CI (0.0076–0.10)
Abdominal wall defect (59)	34	0.77 CI (0.62–0.88)	10	0.23 CI (0.11–0.37)	37	0.88 CI (0.74–0.96)	5	0.12 CI (0.040–0.26)	49	0.91 CI (0.80–0.97)	5	0.093 CI (0.031–0.20)
Other acquired diagnosis (79)	29	0.76 CI (0.60–0.88)	9	0.24 CI (0.11–0.40)	33	0.89 CI (0.75–0.97)	4	0.11 CI (0.030–0.25)	46	0.90 CI (0.79–0.97)	5	0.098 CI (0.033–0.21)

CI = 95% confidence interval
Proportion calculated on patient with results (i.e., denominator adjusted for "no results" in each category)
Table II shows the abnormal coagulation screens as a proportion of all the coagulation screens done for each diagnostic category

Table III: Pearson correlation coefficient and *p*-values

	International normalised ratio	Partial thromboplastin time	Platelet count	C-reactive protein
International normalised ratio	----------	0.3478; $p < 0.05$	-0.3062; $p < 0.05$	0.2223; $p < 0.05$
Partial thromboplastin time		----------	0.1943; $p < 0.05$	0.0978; $\mathbf{p > 0.05}$
Platelet count			----------	-0.2404; $p < 0.05$
C-reactive protein				----------

Table III shows the Pearson correlation coefficients, *r*-values calculated between the different laparotomy results with accompanying *p*-values. Note that the *p*-value for the correlation between CRP and PTT was greater than 0.05, and thus, this *r*-value is not significant

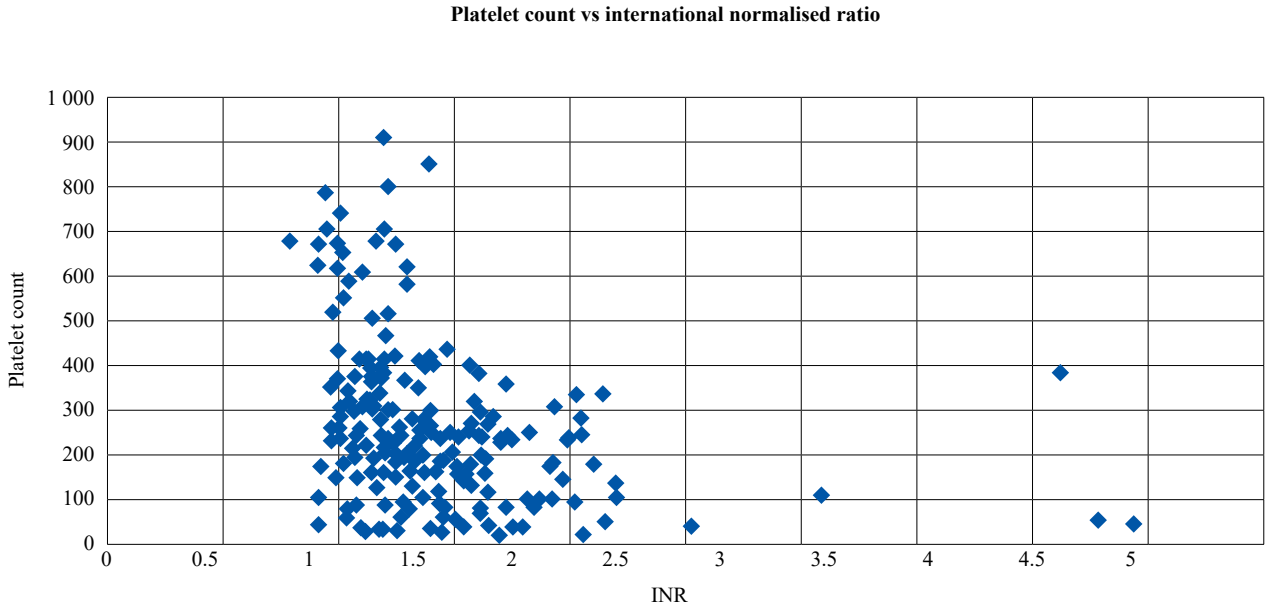

Figure 1: Pearson's correlation coefficient: r = -0.3062 (p < 0.05)
There is a statistically significant moderate negative correlation between platelet count and INR. Of note is that all platelet counts above 400 were associated with a normal INR result of less than 1.5.

the sets of all the results except for the Pearson correlation coefficient between CRP and PTT, where the correlation was not statistically significant. Table III summarises the *r*-values for the pairs of result sets with *p*-values reported.

Result sets with a correlation above 0.3 were assessed further. This included the assessment of the association between INR with platelet count and PTT.

It was noted that above a certain platelet count there were no abnormal INR results. The highest platelet count associated with an INR result higher than 1.5 was 394 x 10^9/L. The relative risk of an abnormal INR in patients with a platelet count above 350 x 10^9/L was 0.2171 (95% CI 0.0831–0.5673). Figure 1 is a scatter chart that shows platelet count and INR results.

Discussion

The understanding of developmental haemostasis suggests that although neonates and infants have different proportions of coagulation factors, they are able to maintain homeostasis under normal circumstances.[12,13] When homeostasis is disrupted in neonates, they are predisposed to disseminated intravascular coagulopathy.[14]

In this study, the researchers compared the rate of coagulopathy in different age classifications, namely *premature neonates*, *term neonates* and *premature infants*. There was no statistically significant difference between the rate of coagulopathy in the different age categories. This suggests that in the setting of a tertiary level hospital in a developing country, where vitamin K is given routinely at birth, gestational age is not an important independent risk factor for coagulopathy in surgical neonates.

When comparing neonates and infants with different underlying diagnoses or reasons for needing to undergo surgery, it was noted that patients with NEC were more likely to have thrombocytopaenia and a deranged INR than other patients.

Sepsis and inflammation are thought to contribute significantly to shifts in homeostasis.[10] CRP was the inflammatory marker assessed in this study. It was expected that there could be a correlation between deranged coagulation markers and raised CRP; however, the correlations calculated between CRP results and other bloods results were weak. The *r*-value calculated was a negative correlation with CRP level against platelet count ($r = -0.24$). This figure represents a weak negative association.

There was a similar, weak association between CRP and INR results. There is no significant correlation between CRP levels and PTT results. These findings suggest that as CRP values increase, higher INR results and lower platelet counts are found. However, the correlation was not compelling enough to guide decision making around omitting preoperative coagulation screening.

There is a moderate negative correlation between INR and platelet count, with an *r*-value of -0.3062. This suggests

that patients with higher platelet counts tend to have lower INR results. The relative risk of an abnormal INR when the platelet count is more than 350 x 10^9/L is 0.2171. This means that a patient with a platelet count of 350 x 10^9/L or more is up to six times less likely to have an abnormal INR than a patient with a platelet count less than 350.

This finding that patients with high platelet counts are associated with normal INR results is clinically useful because many neonatal patients will have a recent full blood count and therefore platelet count at the time of the decision to undertake a surgical intervention. In a resource-restricted environment, with significant delays secondary to laboratory lag times, certain patients with a high platelet count may benefit from omitting coagulation screening and proceeding to surgery without delay if the risk of delayed surgery outweighs the risk of an abnormal INR result.

There was also a moderate correlation between PTT and INR. This correlation was expected given the similarity of the tests.

The retrospective design of this study limited the ability of the researchers to control the timing and techniques of collection of blood samples. Furthermore, not all patients who underwent surgery had blood results. It is expected that a high proportion of the patients who did not have blood results would have been otherwise well. This may have introduced selection bias that could have caused the results to suggest coagulopathy is more prevalent than in reality. Older patients and patients in the age category *premature infants* may have been underrepresented.

All the results calculated in this study were correlations and no comment can be made about the causation of coagulopathy. The researchers did not assess the outcomes in patients who underwent surgery to assess if abnormal blood results were associated with higher rates of transfusion or bleeding.

Conclusion

The rate of abnormal coagulation screen was higher in patients with NEC compared to other surgical diagnoses in the neonatal period. There was no significant difference in the rate of coagulopathy in the different age classifications. It appears that patients with a high platelet count are less likely to have an abnormal INR and that patients with platelet counts above 350 x 10^9/L were up to six times less likely to have a deranged INR than patients with platelet counts less than that cut-off value.

This information can be used in resource-scarce environments to inform decision making around omitting preoperative coagulation screening in select patients where delaying surgical intervention may worsen outcomes.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethical approval was sought and granted by the Health Research Ethics Committee of the study institution affiliated university with ethics number N18/05/054.

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Foetal extraperitoneal rectal perforation in a rural tertiary institution

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Summary
Intraperitoneal rectal perforations have been widely described in the literature, however, foetal extraperitoneal rectal perforations (FERP) remain an extremely rare entity. We describe the twenty-second case contributing to the literature of FERP. The presentation is classic, usually with a perineal and/or buttock deformity evident at birth. Prompt diagnosis and referral is integral in avoiding mortality and morbidity. Complications such as intraperitoneal extension, peritonitis, severe sepsis, and urinary obstruction have been described. The principles of management include initial resuscitation, a de-functioning colostomy, and adequate drainage of the meconium cavity. Prognosis and function are generally excellent with early treatment.
Keywords: foetal extraperitoneal rectal perforation, de-functioning colostomy, meconium cavity

Case report

A 5-day-old, HIV unexposed, term female was born via normal vaginal delivery with a birth weight of 3 250 grams. She was referred to our institution with a 3-day history of progressive left buttock and labia majora swelling, with overlying skin pigmentation and oedema. The patient had been previously fed. Percutaneous aspiration of the cyst demonstrated faecal matter before transfer to our unit. The abdomen had mild distension, was soft and bowel sounds were present. The anus was appropriately positioned and patent. Examination of the mass demonstrated stool draining from the puncture site (Figure 1). A plain abdominal radiograph demonstrated a paucity of rectal air with a non-specific bowel gas pattern. The sacrum was normal and no presacral mass was noted.

Ultrasonography demonstrated a hypoechoic collection with echogenic foci in the left buttock with no extension into the pelvis (Figure 2). The renal system was noted to be normal, however, serology demonstrated acute kidney injury with a urea of 19.1 and a creatinine of 148. The white cell count was 5.17 and the CRP was 251. Based on the above markers, there was a high suspicion of obstructive uropathy and systemic sepsis. An urgent exploratory laparotomy was performed with findings of retroperitoneal meconium staining and swelling evident just above the peritoneal reflection with no intraperitoneal contamination. A significantly dilated bladder was also noted. A divided sigmoid colostomy, drainage of the meconium and faecal cavity through a buttock incision was done and a Penrose silicone drain was left in situ. Broad-spectrum antibiotics were continued postoperatively. Feeds were initiated on the second day postoperatively when the colostomy started functioning.

Figure 1: Significant left buttock and labia swelling with puncture site

Figure 2: Hypoechoic lesion with echogenic foci

Discussion

Foetal extraperitoneal rectal perforation (FERP) is an extremely rare entity that was first described by Mitsudo et al. and coined by Pitcher in 2008. It has a classic clinical presentation as a consequence of antenatal events resulting in rectal perforation.[1-3]

Multiple theories have been postulated regarding its aetiology, however, its exact mechanism remains elusive.[1-3] These include the local ischaemia theory, which is based on the location of the perforation site which is usually in the watershed area between the inferior and middle rectal arteries.[1,2,4] However, this theory is not plausible due to the extensive transmural rectal blood supply.[1,2,4] Another theory is forceful peristalsis against a distal obstruction, which hypothesises that during peristalsis, meconium is forced under pressure through a defect in the lower rectal wall into normal tissue planes within the infralevator space to reach a spinal cord level.[1] After birth, further expansion occurs due to swallowed gas and release of meconium within the tissues with resultant skin rupture.[1] This theory is refuted by the lack of description of distal obstruction in previous studies.[1] A theory proposed by Pitcher et al. suggests herniation of the supralevator rectum through a pelvic floor defect with resultant strangulation and perforation similar to descriptions of Richter's hernias.[1,4]

The most common presentation includes a large perineal, buttock or genital swelling with overlying skin pigmentation secondary to the meconium cavity.[2] The swelling is evident within five days, is often but not always off-midline, soft, non-tender, and may have a fistulous tract draining meconium.[2,5] A normal anus is present in all children.[5] Scrotal enlargement in males can occur due to the extravasation of meconium through a patent processus vaginalis.[6] Delay in diagnosis may result in intraperitoneal involvement complicating with peritonitis, severe sepsis, and a difficult postoperative course.[3-5]

The typical features of this condition do not necessitate special radiological investigations, but, because of its rarity, it may be difficult to diagnose based on clinical findings alone.[2,4,5] Plain radiographs of the abdomen, pelvis, and perineum may demonstrate bowel gas paucity, air-fluid levels or calcifications within the mass or pneumoperitoneum.[2] The diagnostic imaging of choice includes ultrasonography of the perineum, pelvis, and abdomen as well as a contrast study of the rectum.[5] Ultrasonography will display a cavity consisting of echogenic material with anterior rectal displacement and bladder compression.[2,5] Contrast enema is the most common investigation performed and it may demonstrate a rectal perforation or a fistula between the mass and rectum, as well as narrowing and displacement of the rectum.[2,3,6]

Differential diagnoses include rectal duplication, rectal diverticulum, ischiorectal abscess, imperforate anus, and sacrococcygeal teratoma/myelomeningocoele.[1,2,5] A rectal duplication cyst/diverticulum may perforate and have a similar presentation to FERP, however, FERP will not have an epithelial lining histologically.[1]

Recommendations for the management of FERP include faecal diversion through a de-functioning sigmoid colostomy and adequate drainage of the meconium-filled cavity.[2,5] Initial treatment includes resuscitation and broad-spectrum antibiotics especially if the diagnosis is delayed.[4] A de-functioning colostomy should not be deferred if there is an absence of a rectal perforation on contrast enema as this finding may be false negative.[5] Points of contention include the use of contrast enema preoperatively as a diagnostic tool, the need for exploratory laparotomy/laparoscopy, and debridement of the meconium cavity.[2] In the majority of cases, an exploratory laparotomy was performed to rule out intraperitoneal contamination and urinary obstruction.[2,6] Radical debridement of the meconium cavity is not recommended as the collection is extraperitoneal, the meconium is sterile and it poses minimal risk of iatrogenic injury to pelvic nerves and structures.[3,5] Perineal debridement is warranted in the presence of meconium cavity contamination.[2] Stoma closure can be considered 3–6 months after birth and generally spontaneous closure of the rectal perforation would have occurred by this period.[2,3,5] However, a contrast study before the closure of the stoma is warranted as one case had a stricture at the perforation site which subsequently required dilatations before stoma closure.[5]

Rectal biopsies should be performed as part of the workup for distal bowel obstruction to rule out Hirschsprung disease.[2] In one case by Charlton et al., rectal suction biopsy and histological examination of resected bowel excluded Hirschsprung disease by demonstrating ganglion cells and lack of hypertrophic nerve trunks.[2]

Conclusion

Foetal extraperitoneal rectal perforation is an extremely rare condition that has classical clinical features. The presentation is within a few days after birth. Treatment recommendations include stringent resuscitative measures, a de-functioning colostomy, and drainage of the buttock meconium cyst. A rectal biopsy should be performed to rule out Hirschsprung disease as a cause of distal obstruction. This is followed by the closure of the colostomy at 3–6 months after a contrast enema has confirmed distal patency and closure of the fistula.

Conflict of interest

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Getting it straight: hypospadias management in a single centre – the advantages of a uniform approach

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Background: Hypospadias is a common congenital condition in boys, yet the ideal surgical management remains elusive. There are international publications on the topic, but the South African story is largely unknown.
Methods: In this retrospective review, we describe the distribution of disease according to classification, treatment modalities, the outcomes and complications of a single surgeon from Johannesburg, South Africa.
Results: We found distribution of disease (hypospadias type) and complication rates to be similar to those reported in the international literature. Distal hypospadias types account for approximately 70% of disease and are associated with better outcomes, with a complication rate of under 6%, while proximal hypospadias types complicated 50% of the time. Significant risk factors for complications included a delayed time to first operation, proximal hypospadias and those requiring a two-stage procedure. In addition to these risk factors, we describe surgical factors that we feel are crucial in minimising morbidity. These include, but are not limited to, surgery with optical magnification, the use of appropriate fine instrumentation with gentle tissue handling, prioritising the correction of ventral curvature, and adopting a standardised postoperative protocol.
Conclusion: This review offers a relatively small, but important series of hypospadias, demonstrating outcomes equivalent to those achieved in high-volume international centres. We hope it will stimulate further progress in the repair of hypospadias with the aim of further improving outcomes.
Keywords: hypospadias, congenital condition, surgical management

Introduction

Hypospadias is the second most common congenital urological anomaly in newborn males after undescended testes,[1,2] affecting 1 in 200–300 male births.[1,3] The anomaly is characterised by incomplete fusion of the urethral folds during embryogenesis resulting in the urethral meatus being displaced to a more proximal position on the ventral surface of the penis.[3] Associated defects include a variable degree of ventral curvature of the penis, a deficiency of the ventral foreskin, corpora spongiosum and flattening of the glans penis.[4]

Hypospadias is most commonly classified as either distal, where the urethral meatus is located in a glandular, coronal, sub-coronal, distal shaft, or mid shaft position on the penis, or proximal, where the meatus is located on the proximal shaft, or in a penoscrotal or scrotal position.[5,6] Distal hypospadias comprises up to 70% of all hypospadias cases and is associated with better outcomes.[5]

The aetiology of hypospadias is complex and multiple hypotheses have been proposed. These include the presence of a genetic predisposition, inadequate prenatal androgen stimulation, maternal-placental factors, such as maternal hypertension, oligohydramnios and underlying placental insufficiency, as well as environmental influences, including the maternal ingestion of synthetic oestrogens.[7] Ultimately the aetiology is most likely multifactorial.[8]

Hypospadias requires surgical correction in order to improve penile function and provide the patient with a straight penis, with the urethral meatus positioned at the tip of the glans. This will enable the patient to pass urine in a normal standing position, allow for adequate sexual intercourse later in life, and ensure a cosmetically acceptable penis.[6,9,10] Taking into consideration anaesthetic, technical and psycho-development factors, the optimal age for correction is between 6 and 18 months.[4,11,12] Operative repair of hypospadias is one of the most challenging fields in surgery.[1] The fact that over 300 corrective procedures are described in the literature bears witness to the challenges of reconstruction and illustrates the lack of uniformity in surgical approach.[1]

Surgical success rates vary greatly with the weight of complications being carried by patients presenting with proximal hypospadias.[10] The most common complications are urethrocutaneous fistula, glans dehiscence and urethral stenosis.[13] Unsatisfactory cosmetic appearance is often a concern to the family and ultimately the patient, but is not typically listed as a complication.[8] Proximal disease has been associated with complication rates as high as 56% compared to the overall complication rate of 15–22%.[6,14] Proposed explanations for this disparity are based around the associated anatomical defects found with proximal hypospadias. With a more proximal urethral meatus, factors such as a longer neourethral construction, smaller glans, less

penile shaft skin and more severe ventral curvature increase the complexity of surgical repair.[9]

Currently, there is little literature reviewing hypospadias on the African continent and no recent literature reviewing our local experience. This study describes a single surgeon experience in South Africa. We describe the anatomical distribution of disease, time to surgery, type of repair, complication rates and risk factors predisposing to complication.

Material and methods

After ethics approval was attained, convenience sampling was used and all hypospadias patients seen in the single surgeon outpatient clinic from September 2006 to November 2020 were included in this retrospective study. All available clinical (age at presentation and at surgery, hypospadias type, presence and degree of chordee), surgical (number of total surgeries and surgical approaches used) and complication data were extracted from the patient files and recorded in an Excel database. Complications looked for were fistula, glans dehiscence, diverticulum, meatal stenosis, urethral stricture and recurrent ventral curvature. The data were organised, analysed and presented with Stata (Version 16.1 - Statacorp LLC, Texas, USA). Analysis was performed with categorical variables expressed as numbers and percentages. Continuous non-parametric variables were expressed using the median and interquartile range (IQR). Normally distributed variables were described using mean and standard deviation. Inferential statistical tests were used with Fisher exact test for differences in complications for categorical variables and Mann–Whitney U-test for non-parametric continuous variables to test for complications differences between groups (age vs complications). *P*-values of < 0.05 were considered significant.

A uniform surgical approach was used for 96.30% of cases. There are several crucial aspects to our approach, as detailed below.

The patient is placed in the lithotomy position with the surgeon seated comfortably in the midline between the patient's legs. The use of optical magnification is mandatory and a dedicated "hypospadias instrument set" including high-quality fine instruments is essential.[6,12,15] Intraoperatively, correction of ventral curvature is crucial in attaining good surgical outcomes.[6] After applying a tourniquet at the base of the penis to maintain a bloodless field for dissection, an artificial erection is created by injecting saline through the top of the glans into the corpora, and the degree of ventral curvature assessed. The urethral plate is now marked and incisions made laterally, as well as in the midline of the plate, facilitating a tension-free anastomosis. Thereafter a sub-coronal incision is made. For adequate correction of curvature, initially, the entire shaft of the penis must be degloved, this step resolving the vast majority of cases. If the ventral curvature is still present, the fibrous ventral tissue at the apex of the curvature is excised and if necessary, a corporotomy performed. If the penis is still not straight, dorsal plication of the penile tissue can be performed, but not as a substitute for the previously described steps. Once surgical correction of chordee is complete, an artificial erection is again created to confirm that curvature has been corrected. The neourethra is now fashioned using 7/0 vicryl, this over an 8.5 French stent, or a formal catheter in older children who are out of nappies. This urethroplasty is performed in a single continuous layer. On completion of the neourethra, the tourniquet is removed. The glanuloplasty is now performed with absorbable mattress sutures, again using 7/0 vicryl. Next, a midline incision is made in the mobilised foreskin dorsally, defining the Byers Flaps. Where the neourethra extends below the level of the glans, a subcutaneous fascial flap is mobilised and rotated ventrally to cover the neourethra. Finally, the cutaneous Byers Flaps are rotated to achieve ventral skin cover. The urethral stent is sutured in situ, and an occlusive dressing applied. The child is discharged on the day of operation, with a 10-day supply of analgesia, prophylactic antibiotics and oxybutynin to reduce bladder irritation. The dressing and stent are removed in the clinic on postoperative day 10.

Staged repair is indicated in cases where complete correction of ventral curvature cannot be achieved without division of the urethral plate. In these cases, once the plate is divided, the ventral curvature is corrected at the expense of the urethral meatus being moved more proximally, resulting in a significant ventral defect. This defect needs preparation for tubularisation and neourethra creation, which is then carried out at a later second stage.

Results

A total of 162 patients fulfilled the inclusion criteria. Eighty-one patients (50%) presented primarily and had their index surgery in the unit. This cohort of patients form the study population.

Thirty-nine patients (24.07%) were consulted but not operated on for various reasons, and these patients were excluded. Forty-two of the 162 (25.93%) were referred for revision surgery after having their index surgery elsewhere. Thirteen of the 42 were not operated on, while the remaining 29 underwent a secondary surgical intervention.

For the 81 index cases, the median age of presentation was 14 months (IQR: 11, 27) and median age of index surgery was 15 months (IQR: 13, 30). There were three outlying patients with late presentations, presenting at 106, 127 and 156 months respectively. Fifty-two (64.20%) patients presented for the first time before 18 months of age, with 48 (59.26%) of these patients going on to have their index surgery before 18 months. Fifty-seven (70.37%) of the index cases were classified as distal type hypospadias (distal shaft: $n = 42$ and midshaft: $n = 15$), while 24 (29.63%) cases were proximal type (proximal shaft: $n = 6$, penoscrotal: $n = 17$, scrotal: $n = 1$). Thirteen (16%) patients with hypospadias did not have any ventral curvature. Twelve of these 13 cases were associated with distal hypospadias and one with midshaft hypospadias. Of the 84% of cases with curvature, 11.11% ($n = 9$) were classified as mild, 45.68% ($n = 37$) moderate and 43.21% ($n = 35$) as severe.

Although there is poor conformity in surgical approach to hypospadias repair,[1] the majority (96.30%) of patients in this study were managed with a uniform approach, either undergoing a single-stage tubularised incised plate (TIP) procedure or a staged repair.

Sixty-two of the 81 (76.54%) index cases had a single-stage TIP procedure. Thirty-nine (62.90%) of these procedures were performed in cases of distal hypospadias, 15 (24.19%) in midshaft cases and the remaining 12 (38.7%) in cases of more proximal hypospadias. Early in the experience, three meatal advancement glanuloplasty incorporated (MAGPI) procedures were performed. They were all performed in cases of distal hypospadias and resulted in no complications.

Table I: Descriptive statistics of hypospadias types (n = 81)

Hypospadias type	n (%)	Tubularised incised plate procedure (%)	Two-stage procedure (%)	Postoperative complications (%)
Distal	42 (58.85)	39 (92.86)	0 (0)	3 (7.14)
Midshaft	15 (18.52)	15 (100)	0 (0)	0 (0)
Proximal	6 (7.4)	4 (66.67)	2 (33.33)	2 (33.33)
Penoscrotal	17 (20.99)	5 (29.41)	12 (70.59)	10 (58.82)
Scrotal	1 (1.23)	0 (0)	1 (100)	1 (100)
Total	**81 (100)**	**63 (77.78)**	**15 (18.52)**	**16 (19.75)**

Fifteen of the 81 (18.52%) cases underwent a two-stage procedure. All 15 of the staged repairs were performed for proximal types (proximal shaft: n = 2, penoscrotal: n = 12, scrotal: n = 1).

Of the 81 index cases, 17 (20.99%) developed postoperative complications. Urethrocutaneous fistulae accounted for 16 (19.75%) of these. Only one (1.23%) urethral stricture occurred. Of the 17 cases that had complications, 14 occurred in patients with severe ventral curvature (82.35%). Fistulae developed in three of the 57 (5.26%) cases of distal type hypospadias (distal: n = 3; midshaft: n = 0), while 12 of the 24 (50%) cases of proximal hypospadias developed fistulae (proximal: n = 2; penoscrotal: n = 9; scrotal: n = 1). Complications were noted to be significantly higher in the following patients: those with proximal hypospadias compared to more distal types ($p < 0.001$), patients who underwent two-stage procedures ($p < 0.001$) and those who were at a delayed age at the time of first surgery ($p = 0.0066$).

Discussion

There is no consensus on surgical technique for hypospadias repair with operative approach and outcomes varying significantly.[1,4,6] In addition, there is a paucity of African data on the topic. Hypospadias is commonly diagnosed at birth,[6] however, the median age of initial presentation in our cohort was 14 months and for index operation 15 months. Current recommendations suggest that the optimal time for hypospadias repair is between 6 and 18 months of life. At this age, anaesthetic, anatomical and psycho-developmental factors are optimised.[4,11] Of our 81 patients who presented for index surgery, 48 (59.26%) were operated on before 18 months of age. The specific reasons for later presentation and hence operative repair are currently unknown but may be due to a lack of public awareness of an often-stigmatised anatomical disorder as well as inappropriately late referral from medical specialists unfamiliar with this disorder. Consistent with the described anatomical distribution of hypospadias type,[5] the majority (70.37%) of index cases were classified as distal with the remainder being proximal. Ventral curvature was present in 84% of all index cases.

All 81 patients presenting for index operation to our unit had one of three operations performed: a single-stage TIP repair, a staged repair or a MAGPI repair. The three MAGPI procedures were performed for cases of glanular hypospadias with megameatus early in our experience, but the TIP procedure is now used to repair this category of hypospadias too. The repair of hypospadias is regarded as one of the most challenging fields in surgery and this is borne out in the high and variable complication rates seen across the world. An overall complication rate, across all types of repairs, of 15–22% is frequently reported.[16] In the African context, recent literature includes a single centre review of Nigerian hypospadias patients published in 2015 and a 2020 Tanzanian paper looking at local hypospadias surgery outcomes. The Nigerian paper described an overall complication rate of 50%,[17] while the Tanzanian paper described an overall complication rate of 61%,[18] both much higher than the internationally recognised rates of 15–22%.[16,17] The complication rates for proximal hypospadias repairs, which are classically higher than the overall rates, were 88% in the patients from Tanzania,[18] but were not stated in the case of the Nigerian paper. This series reports an overall complication rate of 20.99%, which is in keeping with those reported from high-volume international centres, and significantly better than the outcomes currently reported from the African continent.[11,16,19-21]

Distal hypospadias repair is known to complicate less frequently than repair of proximal hypospadias, with expected complication rates for distal type hypospadias documented at between 5–10%.[6] In a 2010 systematic review of 28 papers looking at complications of distal type hypospadias, an overall complication rate of 5.52% (186/3 367) was found.[19] The majority (67.20%) of these complications were fistulae. Similarly, in a 2013 comparison of seven published papers on distal hypospadias repair, an overall complication rate of 7.32% (71/970) was noted.[11] Forty-one of the 71 (57.75%) complications were fistulae.[11] We report an equivalent complication rate of 5.26% in our cohort of distal type hypospadias patients. All of these were fistulae.

Penile anatomy in proximal hypospadias is frequently more distorted than that in distal hypospadias. Salient differences include a relatively smaller glans penis, relatively less penile shaft skin available for reconstruction, as well as more severe ventral curvature.[20] These factors increase the complexity of surgical repair and increase the incidence of complications.[2,10] Complication rates for proximal hypospadias with severe curvature are reported to be between 22–53%.[6,10,16] Recently, Boston Children's Hospital reported a complication rate of 52.99% (71/134), while Texas Children's Hospital had a rate of 67.86% (38/56),[20,21] and Chung et al. described complications in 48.89% of patients (22/45).[14] Whilst a relatively small cohort, our cases of proximal type hypospadias followed the expected pattern of having significantly higher postoperative complication rates than distal type hypospadias ($p < 0.001$). Fifty per cent of patients post proximal hypospadias repair developed fistulae, while one patient developed a urethral stricture (4.17%).

We found risk factors for development of complications included delayed age of first surgery ($p = 0.0052$), proximal types of hypospadias ($p < 0.001$) and those requiring a two-stage procedure ($p < 0.001$). In attaining the above

acceptable and reproducible results, adherence to several critical surgical factors is felt to be mandatory.

The surgeon should be seated comfortably in the midline and make use of optical magnification and high-quality, fine instruments. A bloodless field is maintained by placing a tourniquet at the base of the penis. Correction of ventral curvature is crucial, with most cases being resolved by penile degloving only. An artificial erection must be used to initially assess the degree of curvature and again post correction to ensure that the curvature is adequately corrected. All anastomoses must be tension free while the neourethra is fashioned over an 8.5 French stent using 7/0 vicryl. A standardised postoperative protocol should be used. Leaving a urethral stent in situ for 10 days and prescribing oxybutynin and prophylactic antibiotics should form part of this protocol.

Conclusion

Hypospadias is a common congenital condition in boys, yet the ideal surgical management remains elusive. There are numerous international publications on the topic, but the African story is largely unknown. In this review, we describe the pattern of disease, treatment modalities and outcomes of a single surgeon. Distribution of disease (hypospadias type) and complication rates are similar to those reported in the international literature. Distal hypospadias types account for approximately 70% of disease and are associated with better outcomes, with a complication rate of under 6%, while proximal hypospadias types complicated 50% of the time, with the majority of these complications being urethrocutaneous fistulae. Significant risk factors for complications included a delayed time to first operation, proximal hypospadias and those requiring a two-stage procedure. However, over and above these risk factors, we feel that the described surgical factors are crucial in minimising morbidity. These include, but are not limited to, surgery with optical magnification, the use of appropriate fine instrumentation with gentle tissue handling, prioritising the correction of ventral curvature, and adopting a standardised postoperative protocol, including a urethral stent in situ for 10 days, oxybutynin and prophylactic antibiotics. This review offers a relatively small, but important series of hypospadias, demonstrating outcomes equivalent to those achieved in high volume international centres. We hope it will stimulate further progress in the repair of hypospadias, encourage analysis and publication of results with the aim of further improving outcomes.

Study limitations

This study's limitations include its retrospective study design, its convenience sampling as well as being a single centre, single surgeon, review.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethical approval was obtained from the University of the Witwatersrand Human Research Ethics Committee (Ethics Certificate Number M200602).

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PAEDIATRIC SURGERY

What is the functional and cosmetic outcome of the ventral slit procedure for congenital megaprepuce?

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Background: Congenital megaprepuce is a urological condition characterised by a megapreputial reservoir with a long redundant inner prepuce. An impeded urinary stream and resultant ballooning is associated with discomfort and causes parental anxiety due to the unusual appearance of the phallus. Surgical correction should improve functionality, but cosmesis is also important. This study took place in a community in which traditional circumcision remains an essential rite of passage. Therefore, early surgical correction of congenital megaprepuce was complicated by the unique requirement that patients remain uncircumcised. The aim of this study was to assess the functional and cosmetic outcome of the ventral slit procedure (an uncomplicated technique used to restore urinary flow which, importantly, preserves the foreskin), as reported by the parents of the patients.
Methods: Here we investigated the functional and cosmetic outcomes of the ventral slit procedure, in a retrospective review of patients operated on between 2014 and 2018 in our hospital – Red Cross War Memorial Children's Hospital. Twenty-five cases were identified, of which only 18 patients were contactable telephonically. Parents of 18 paediatric patients were interviewed postoperatively in a telephonic survey regarding phallic appearance and functionality following surgery.
Results: Overall, the ventral slit procedure successfully restored flow, prevented ballooning and alleviated discomfort during voiding in all 18 patients. Parents interviewed were highly satisfied with surgical outcomes, as assessed by the paediatric penile perception score.
Conclusion: The ventral slit procedure was found to be a culturally acceptable and simple surgical solution to congenital megaprepuce.
Keywords: urology, genitourinary surgery, congenital megaprepuce, ventral slit procedure

Introduction

Congenital megaprepuce (CMP) is a penile malformation characterised by a megapreputial reservoir with a long redundant inner prepuce. The condition is often associated with problematic urination, frequent urinary tract symptoms and urinary tract infections (UTIs), as well as parental concerns about phallic size due to ballooning of the penis during voiding. O'Brien et al.[1] first described CMP in 1994, and since then many surgical options have been described.[2-9] The natural history and evolution of this CMP is unclear, and it may be the result of a proximal stenotic preputial opening which in turn creates the impression of a foreshortened penis with ballooning of the megaprepuce during micturition (Figure 1), and urine leaking from this megapreputial reservoir.

By definition, the condition should be present at birth, however it is often overlooked. Moreover, diagnosis of CMP should be distinguished from several other anomalies such as buried penis – a common anomaly frequently associated with insufficient outer penile skin, inadequate subcutaneous attachment to Buck's fascia and usually a narrow opening of

Figure 1: Typical appearance of a congenital megaprepuce

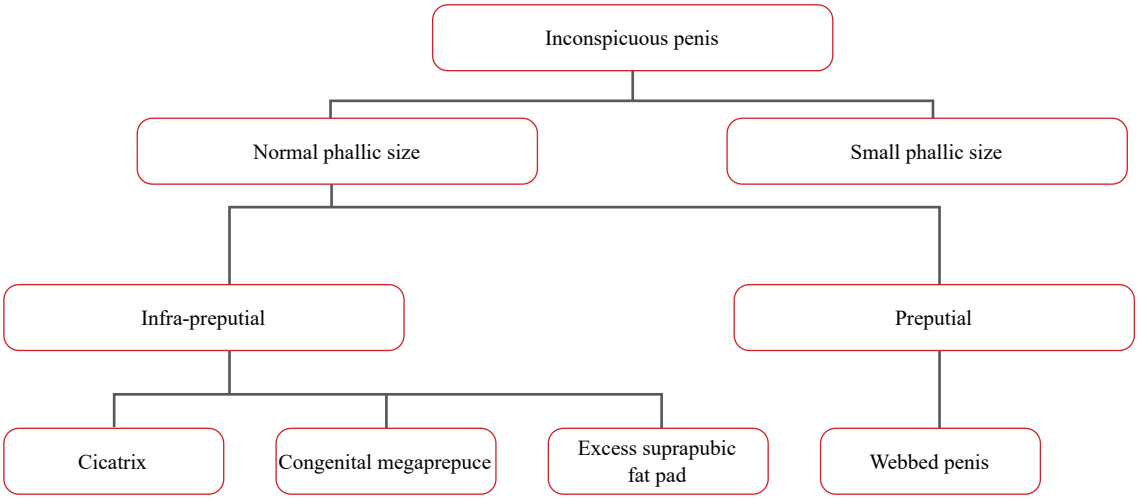

Figure 2: Schematic demonstrating the criteria described by Alexander et al. for diagnosis of CMP

the prepuce; trapped penis – a penis of normal size that has regressed behind an iatrogenic post circumcision cicatrix; webbed penis – congenital condition in which the scrotal skin extends onto the ventral penile shaft, tethering the shaft to the scrotal midline by a skin web; micropenis – an unusually small penis that is normal in appearance but less than two standard deviations in size when erect or stretched for the mean age group; and concealed penis – a penis hidden by an excessive suprapubic fat pad and poor fixation of the shaft skin.

Several workable classifications have been used to describe the pathological anatomy. Alexander et al.'s classification[2] is most helpful in the clinical setting when faced with a child with an "inconspicuous penis". The logic flows from determining the phallic size (usually normal in this context), then the glanular position, a clear description of the condition (trapped penis, buried penis, concealed penis and webbed penis), and the underlying cause (post circumcision cicatrix, congenital megaprepuce, excessive suprapubic fat pad and penoscrotal fusion)[2] (Figure 2). Alternative classification schemes include Maizels et al.'s classification of the concealed penis,[8] Crawford's,[9] and Hadidi's graded classification system, in which CMP falls under grade 1.[10]

With regard to surgical correction of CMP, the objectives are to restore the functional anatomy with an unobstructed urinary stream and to improve the cosmesis. Moreover, corrective procedures should ideally be performed early in life. Several surgical techniques have been described including multiple Z-plasty, removal of the suprapubic fat pad, the use of preputial island flaps, fixation of the suprapubic skin to the pubis, preputial unfurling, ventral V-plasty, the double longitudinal megapreputium incision technique (DOLOMITE) technique and various degloving/penoplasty techniques.[2-7,10-15] These procedures are all complex procedures requiring considerable levels of surgical skill. All of them result in the circumcised appearance of the penis.

The ventral V-slit was first described in La Vega, as a treatment for phimosis on an island where circumcision is considered culturally unacceptable. It is a simple procedure where once an adequate level of anaesthesia of the patient is achieved (general anaesthetic with a penile ring block using Bupivacaine), a straight haemostatic clip is placed ventrally

Figure 3: Triangulation with holding sutures, and crushing haemostat in midline

Figure 4: Ventral slit performed

Figure 5: Mikulicz suturing of skin edges completed, and appearance with foreskin retracted reveals glans and inner preputial folds

in the midline of the prepuce for half a minute (Figure 3). Tissue scissors are then used to dissect along this line of crushed tissue, which in turn allows the glans to be exposed as the prepuce retracts. The incision is made to extend beyond the phimotic narrowing (Figure 4). Holding sutures can be used to help triangulate the incised prepuce into a V-shaped pattern, and then the skin edges are approximated with a 5/0 chromic running suture (Figure 5).

In comparison to all other published techniques, the ventral slit procedure to achieve an unobstructed urinary system is simple and results in an uncircumcised appearance of the phallus.[13]

Setting

At Red Cross War Memorial Children's Hospital (RCWMCH), Cape Town, South Africa, the majority of our patients with congenital megaprepuce are Xhosa speaking.

The community surrounding the hospital are largely Xhosa, in which traditional circumcision, known as Ulwaluko, plays an extremely important socio-sexual role in the transition from childhood to manhood.[16] The initiation ritual traditionally takes place around the time of physical puberty; therefore it is a unique requirement of this region that any surgical correction for CMP leave the patient uncircumcised in order to preserve future rites of passage. We therefore began to think of surgical options for congenital megaprepuce that were foreskin preserving and started to perform the ventral slit procedure. At Red Cross we also offer patients the Ventral V-plasty technique and a novel inverted circumcision technique to those patients who want a circumcised appearance of the foreskin for CMP.[2]

Table I: Parental responses to the buried penis questionnaire

Question	No (n = 18)	Yes (n = 18)	If yes, please comment
Did your child have infections prior to surgery for buried penis?	15	3	2 patients reported UTI
Did he develop an infection after surgery?	16	2	2 patients developed postoperative wound infections, but no UTI reported
Did urine pool behind skin prior to surgery for buried penis?	0	18	All parents reported ballooning of foreskin
Did urine pool behind the skin after surgery?	17	1	1 patient experienced postoperative wound problems and required further surgery
Did you have trouble keeping your son's skin clean prior to surgery?	18	0	
Do you have trouble keeping your son's skin clean now?	18	0	
Was the penis completely hidden behind skin prior to surgery?	0	18	
Is the penis hidden after surgery?	17	1	The patient who had wound infection postoperatively had an issue with concealment
Did your son experience pain with voiding before surgery?	6	12	
Does your son experience pain with voiding after surgery?	18	0	
Did you have negative concerns about the appearance of your son's penis prior to surgery?	0	18	
Do you think the surgery increased the length of your child's penis?	14	4	
Do you think surgery was helpful in alleviating those negative concerns?	0	18	
Would you want this surgery again or recommend it to a friend whose child had the same condition as your son?	0	18	
Did surgery make your son's penis more assessable?	0	18	

Table II: Cosmetic assessment of penis following the ventral slit procedure

GPS	Very satisfied (n = 18)	Satisfied (n = 18)	Dissatisfied (n = 18)	Very dissatisfied (n = 18)
Meatal position and shape	18	0	0	0
Shape of the glans	17	1	0	0
Shape of the penile skin	16	2	0	0
General cosmetic appearance	14	4	0	0

Aim

The aim of this study was to evaluate whether the functional and cosmetic outcome of the ventral slit procedure is acceptable for CMP, in a culture that does not accept circumcision before a certain age as a rite of passage into manhood.

Methods

We performed a retrospective review of the urological database at the RCWMCH from 2014 to 2018 of all male infants who had a ventral slit for CMP. Participants were then contacted telephonically and invited to attend an outpatient appointment or telephonic interview to prospectively collect outcomes data. The study included both an interview with parents or legal guardians as well as an intended clinical examination conducted postoperatively. Parents and guardians of study participants were asked to complete the buried penis questionnaire (Table I), which was used to assess functional and cosmetic outcomes. The paediatric penile perception score (Table II) was then used to report on phallic appearance where meatus, glans, penile skin and overall cosmetic appearance were scored. Informed consent for each participant was granted by parent or legal guardian. The study was a prospective observational review of male infants who have had the ventral slit procedure for CMP at the RCWMCH between 2014 and 2018.

Results

Initial observations

A total of 25 cases were identified, of which 18 were contactable. Eighteen male infants who had the ventral slit procedure for CMP performed at the RCWMCH were therefore included in the study. Seventeen infants included were South African males of Xhosa descent. Seventeen participants opted for a telephonic interview, whilst one patient attended an outpatient appointment. The mean age of participants at the time of procedure was 13 months (range 4–31 months). All participants underwent the surgery under general anaesthetic, with two-thirds of the patients managed as day-case surgery. Mean operative time was 29 minutes (range: 7–60 minutes), with the mean anaesthetic time of 61 minutes (range: 30 minutes to 2 hours). The child who required a surgical time of 60 minutes, desaturated intraoperatively, and required a second procedure by an ENT surgeon simultaneously to drain a vallecular cyst. Two patients had issues with wound dehiscence postoperatively and stayed in hospital longer than a day. Three patients underwent further surgery: one patient required epispadias repair (incidentally detected at the time of surgery), one patient developed paraphimosis postoperatively and underwent formal circumcision, and another patient opted for further surgery with the circumcised appearance as the outcome.

Buried penis questionnaire

The functional outcomes were as follows, using a binomial distribution for the p-values:

- There was a 17% drop in UTIs (p = 0.00067)
- None of the patients had ballooning of the foreskin
- Only one patient had hidden foreskin out of 18 (p = 0.014)
- No patients experienced dysuria postoperatively
- All 18 would recommend surgery to a friend

The paediatric penile perception score

The paediatric penile perception score (Table II) was used to evaluate penis cosmesis following the ventral slit procedure. One participant was available for postoperative examination, in this case assessment was made by the author. In all other cases the paediatric penile perception score was used to assess contentment of the parent or guardian. All the patients were too young to answer the questionnaire themselves.

The single patient who was clinically evaluated was 4 years old at the time of review, having undergone surgery at the age of 10 months. He had a normal length phallus, with a good penoscrotal angle. He had no visible scar tissue, and no tethering of skin with the penile shaft. The foreskin appeared normal for a child his age and retracted easily.

Discussion

This study draws attention to the simplicity of the ventral slit procedure as a suitable corrective procedure for CMP. The procedure is technically uncomplicated to perform, has minimal postoperative sequelae, and offers the additional advantage of leaving the foreskin intact.

Despite the small numbers, we were able to demonstrate a statistically improved absence of dysuria and ballooning of the foreskin and a non-significant trend toward decreasing UTIs.

Despite the small numbers we were also able to demonstrate a statistically significant cosmetic satisfaction score among parents.

As indicated in the buried penis questionnaire, the procedure successfully alleviated pathology associated with CMP, and relieved discomfort during voiding. Ballooning of the penis due to pooling of urine behind the foreskin, which was noted as a particular source of parental anxiety prior to surgery, was assessed in every case. Importantly, all parents and guardians found the ventral slit procedure to be an acceptable surgical correction for CMP and were mostly "very satisfied" with phallic appearance post-surgery. If further surgery was desirable to leave the child with a circumcised appearance, this could be performed at a later stage when the child is of a consenting age. In this

case, surgery would also be less technically challenging as the child would have outgrown any issues associated with this condition if left unchecked, namely the remarkably redundant inner prepuce and the associated psychological embarrassment associated with a concealed penis.

Extensive waiting lists for surgery delay corrective measures. Ideally, these patients should be operated on as soon as possible after the diagnosis is made, as this would alleviate discomfort associated with urination and parental concerns. Moreover, the correction of the abnormality before the child becomes self-aware could avoid significant humiliation in the school environment.

Not only does the ventral slit procedure make for a culturally appropriate choice for surgical correction, but it could mitigate surgical delays as the operation itself is not technically difficult. The procedure itself is also not time consuming – our mean operative time being 29 minutes and mean anaesthetic time of 61 minutes. Patients could be prioritised for day-case surgery which would increase the number of patients treated per day.

The limitations of this study are that none of the patients had undergone traditional circumcision at the time of the study, and it would be useful to understand whether any issues arise in the population study group. Only one patient had outpatient follow-up, due to the restrictions of the COVID-19 pandemic on routine follow-up or research in the outpatient setting.

Our recommendations for future research would be to compare circumcising techniques with the ventral slit procedure, as we feel mean operative times would be statistically different, whilst cosmetic and functional outcomes could rival each other. It would also be useful to validate this procedure in other settings where there was not necessarily an emphasis on whether the child appeared circumcised or not after surgery.

Conclusion

To the best of our knowledge this is the first description of ventral slit for CMP. We were able to demonstrate that it gives a good functional and cosmetic outcome to those patients with CMP requesting foreskin preservation. The authors therefore recommend the ventral slit procedure as a suitable operation to restore flow dynamics for young male infants suffering from CMP. In this region, it represents a culturally acceptable surgical solution, with favourable cosmetic outcomes and could also bring relief to our currently overburdened surgery waiting lists.

Conflict of interest

The authors declare no conflict of interest.

Ethical approval

The study protocol was approved by the UCT Health Science Human Research Ethics committee, ethics number 124/2019.

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From Canada to South Africa – the use of artificial intelligence to assist with management decisions in children with hydronephrosis

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Background: Paediatric hydronephrosis (HN) often resolves spontaneously, however some patients with obstructive HN may require surgery to minimise long-term sequelae. Artificial intelligence (AI) could help predict the likelihood of obstructive HN using ultrasound images. This study explores clinician attitudes towards the use of such a tool, developed by a paediatric urology unit in Canada, in their own practice in South Africa (SA).
Methods: Doctors dealing with HN in day-to-day practice were approached to participate. Participants completed a standardised questionnaire, followed by an interview. Here, they were presented with structured questions based on survey responses, as well as unstructured topic areas to explore.
Results: Twenty-three doctors across seven provinces were interviewed, representing specialities including paediatric surgery, urology, general paediatrics, nephrology and radiology. Doctors of all types were open to the use of AI tools to assist with decision-making in children with HN, provided the tool is validated in SA. Discussion themes included concerns around input-image quality and difficulties with imaging access. Several doctors expressed an interest in AI tools that could assist them to better perform their own point-of-care ultrasound (POCUS) and then provide some image analysis, as well as a digital application that allowed the input of various health data points to create a comprehensive referral package for individual nephro-urological patients.
Conclusion: A validated digital tool that incorporates AI to assist with management decisions in children with HN would be potentially welcomed in SA. Barriers include access to high-quality paediatric ultrasound (US) imaging and clinician willingness to adopt new technologies.
Keywords: paediatrics, hydronephrosis, artificial intelligence, machine learning, uteropelvic junction obstruction

Background

Chronic kidney disease (CKD) is a common and costly health condition, a risk factor for cardiovascular disease, as well as a precursor to end-stage renal disease (ESRD).[1-3] Childhood kidney disease has been found to be closely linked with CKD and ESRD in adult life.[2,4] In many cases, advanced CKD can be prevented or mitigated with proactive medical and clinical intervention, especially in childhood. This is possible by close observation, regular monitoring and early intervention when needed.[5,6] Herein, we seek to understand the barriers to and opportunities for developing an ultrasound (US) image-based artificial intelligence (AI) tool to detect and guide the treatment of a common childhood kidney anomaly, hydronephrosis (HN), within the South African healthcare system.

HN is a common prenatal ultrasound finding (1–5% of foetuses) but outcomes for these patients vary widely.[7,8] The majority (70%) of HN resolves without intervention, but medical and/or surgical intervention is indicated to avoid long-term renal damage in patients with an obstructive aetiology such as ureteropelvic junction obstruction (UPJO).[9] In South Africa (SA), the prevalence of paediatric HN is not known, however the incidence of CKD in South African adults is between 15 and 25%,[10] approximately double that of Canada. Because SA is a mixed low- and middle-income country (LMIC), with a two-tier (public/private) healthcare system, services such as comprehensive antenatal care are unevenly distributed and access to this care can be difficult, particularly amongst low-income patients.[1,11-14]

In this environment, HN may only be diagnosed once it becomes symptomatic and there is irreversible functional renal damage.[10] Enabling earlier detection and treatment could potentially result in improved overall renal outcomes for the population. Our collaborative research group has already developed a model to predict obstructive HN based on US images alone for the Canadian context.[15] This

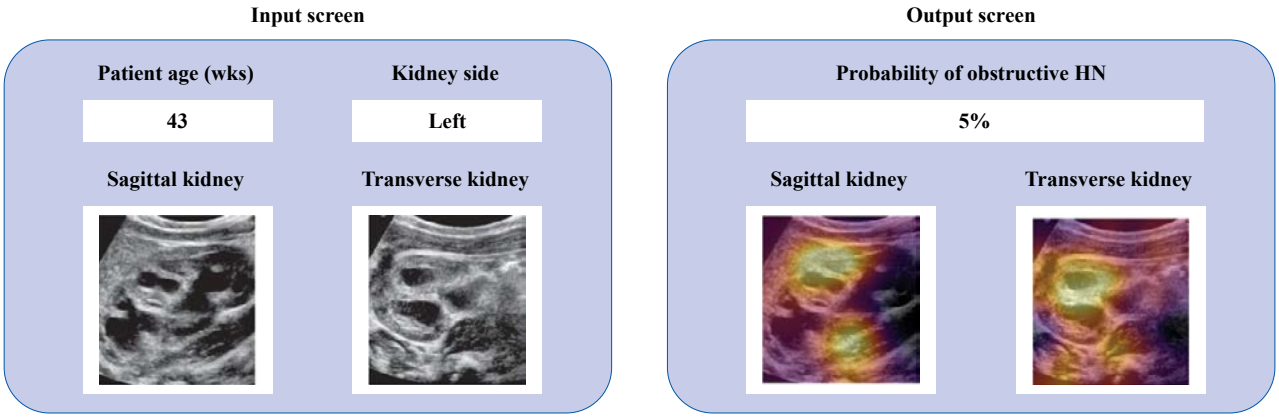

Figure 1: Mock-up of the user interface for an AI application designed to predict the likelihood of obstructive hydronephrosis (HN) based on US images. The 'Input' screen allows the user to upload images and the patient's gestational age and kidney laterality. The 'Output' screen provides a percentage prediction of the likelihood of obstructive hydronephrosis, overlaying a heat-map on the images to allow the user to see which parts of the images were used to make the prediction.

tool, which is still undergoing investigation, examines a kidney US at a single time-point and then makes a prediction regarding the likelihood of obstruction within the hydronephrotic kidney. We have shown that using this tool can reduce invasive testing and follow-up frequency in more than half of patients with non-obstructive HN, without reducing the standard of care for patients with obstructive HN. We hypothesise that a model of this kind could be adapted and used to enable improved HN detection and care in the SA context.[16] A mock-up of the user interface of this tool is shown in Figure 1.

In this exploratory work, we examine how early renal anomalies and dysfunction are identified and treated in the SA healthcare system by interviewing 23 clinicians in varied locations, specialties and career stages. We also explore attitudes towards and perceived challenges around using a Canadian-developed AI tool in the management of paediatric HN in SA. We attempt to establish where opportunities may exist to enhance the use of existing resources and facilitate higher quality care without creating an additional burden for patients, clinicians or the healthcare system. We find important differences in clinical management between the Canadian and South African context. Assumptions in our original HN model based on its development in the Canadian context become clear in this analysis, and opportunities for modifications to the existing tool as well as additional models and technology that have the potential to improve paediatric care are identified.

Methods

Clinicians who would be potentially involved in the management of paediatric HN around SA were approached to participate in the study. For the first round of engagement, clinicians with whom the South African researchers already have an established professional relationship with the authors of this study were approached directly to participate in the process. These 'first-round responders' were each asked to identify one or two potential participants not known personally to the primary researchers who were then approached telephonically or via email to participate. As such, a snowball sampling approach was undertaken to reach the goal number of participants.[17]

Following completion of an informed consent form, participants were asked to complete a REDCap survey to determine participants' demographic details (Table I), to achieve an understanding of their clinical setting and experience, and to get an idea of how frequently and in what role they would interact with patients with HN. Questionnaire responses were then compiled into a spreadsheet and analysed in R v4.0.2 to assess the distribution of participant characteristics.[18] This was followed by an open-ended interview structured to cover various topic areas, including how patients with HN are identified and treated in the interviewees' settings, where technology currently fits into the clinical setting, and viewpoints on AI technology in this context (Appendix A). We recorded and then transcribed the content automatically using Microsoft Office tools, all while being stored within the hospital firewall. Interviews were then coded over two rounds to generate themes for our results.[19,20]

Results and discussion

Study participants

We interviewed 23 doctors across seven provinces. Our participants were between the age of 26 and 55, with eight medical officers, one postgraduate trainee, and 14 specialists. These clinicians were working in the Western Cape (n = 4), Gauteng (n = 9), the Free State (n = 2), the Eastern Cape (n = 5), KwaZulu-Natal (n = 1), the Northern Cape (n = 1) and North West Province (n = 1). Respondents were asked to identify which areas they worked in (they could pick more than one). The field most represented was paediatric surgery (n = 9), followed by general medicine, general paediatrics and nephrology (n = 4 each), urology and neonatology (n = 3 each), radiology (n = 2), and foetal medicine and obstetrics (n = 1 each). Doctors working in the state sector (n = 21), the private sector (n = 9) and academic practice (n = 15) were represented. Three quarters of respondents were based at a hospital 50 km or less from their nearest referral unit.

Detection and management of hydronephrosis

Throughout the interviews, important differences in the clinical presentation of HN in SA versus Canada, where this tool was developed, were highlighted. In both countries, postnatal renal US in asymptomatic patients is most often performed following an irregular finding on a prenatal US.

Table I: Demographic details of participants

Age	25–55 years (mean = 38)
	n (%)
Province of work	
Gauteng	9 (39)
Western Cape	4 (17)
Eastern Cape	5 (22)
Free State	2 (9)
KwaZulu-Natal	1 (4)
North West	1 (4)
Northern Cape	1 (4)
Facility level*	
Quaternary	11 (48)
Tertiary	16 (70)
Secondary	3 (13)
District/primary	1 (4)
Distance to nearest referral centre	
0–50 km	17 (74)
50–100 km	1 (4)
100–300 km	2 (9)
300–500 km	1 (4)
> 500 km	2 (9)
Current position	
Specialist	14 (61)
Postgraduate trainee	1 (4)
Medical officer	8 (35)
Current field*	
Paediatric surgery	9 (39)
Urology	3 (13)
Nephrology	4 (17)
General paediatrics	4 (17)
Neonatology	3 (13)
General medicine	4 (17)
Radiology	2 (9)
Obstetrics	1 (4)
Foetal medicine	1 (4)
Primary role in paediatric HN*	
Diagnosis	17 (74)
Referral	14 (61)
Expectant or medical management	14 (61)
Surgical repair	9 (39)

* participants were able to choose more than one option

Otherwise, it tends to be performed after repeated urinary tract infections (UTIs). In SA, prenatal US is often either unavailable or not reviewed by a specialist. Likewise, UTIs may often go undetected, as fevers may often be treated with antibiotics without the underlying cause being thoroughly investigated. This phenomenon may be driven by the difficulty in obtaining a non-contaminated urine sample from very young children to verify the UTI using a urine dipstick.[21] Additionally, young patients can often not describe their symptoms verbally. Thus, indicators of early renal disease (repeated UTIs and anomalies visible via imaging) may persist for a great deal of time without detection or adequate treatment. Early renal disease is thus relatively "quiet", with subtle features including poor growth and developmental delay being easily missed. Several specialists pointed to this as a problem leading to lower awareness of renal disease relative to more profoundly symptomatic cardiac or neurological issues which have a far more dramatic clinical presentation. One specialist described this issue by saying 'We should have our [blood pressure] and a urine dipstick once a year, the month of your birthday so we don't forget, and that way you'll pick up lots of renal issues because renal is a silent killer. It's a complete nightmare. If you're cardiac, you'll be blue. If you've got epilepsy, you'll be fitting. But the poor old renal kids, they're just cute, they're short, they have a [blood pressure] that's high that no one's ever going to pick up, and they look slightly pale.'

Because US is often not performed on high-risk patients in infancy, our initial model built with early US images would likely require revision to accommodate older patients. Multiple participants stated that awareness of renal disease and accessibility to expertise (easy to access clinical recommendations or correspondence with a specialist) would be valuable in any application built to assess paediatric US (for HN and beyond).

Ultrasound imaging and interpretation

A concern about poor US imaging quality was raised numerous times as a potential barrier to using something similar to our original tool which relies entirely on US images of the kidney. Even quaternary centres may not have specially trained sonographers on staff to perform renal US in paediatric patients. One radiologist interviewed stated that US is the least preferred imaging modality from the perspective of radiologists and is therefore often left for the most junior radiologists to perform, often without much formal training around the modality. US is always user-dependent, and paediatric renal US requires the user to understand that there are some fundamental differences in the appearance of paediatric vs adult kidneys, which may not be well appreciated by radiologists who do not report paediatric US on a regular basis.

Similar to Canada, the clinicians we interviewed who worked at the secondary, tertiary and quaternary centres in SA all used picture archive and communication systems (PACS) to digitally store and access USs. Doctors from one tertiary centre described their ability to view and evaluate ultrasounds from multiple secondary centres which refer to them. However, many interviewees indicated that they rarely view US images and tend to rely heavily on the content of reports for their decision-making. This was due to a combination of junior doctors having very little experience in interpreting US imaging, as well as difficulty accessing images. Several interviewees who worked at a centre with PACS stated that there were very few places in the hospital that allowed the viewing of images. At the primary level USs are often not connected to any kind of storage system and are printed and affixed to a report describing the US which is given either to the clinician or patient – both of which make follow-up and recovery of images challenging. In the worst-case scenario, many sites run out of paper to make physical copies of images on a regular basis, thus leaving no record of these images beyond the written report following the scan. Concerns around the accuracy of reports were also raised: one radiologist interviewed said "I think that there's

a shocking amount of radiology reports that go out [with] mistakes." The accuracy of the report relies on the accuracy of the sonography, both in terms of their sonographic skill and in terms of their dictation.

Given these challenges, several specialists interviewed suggested that an AI tool would need an algorithm to provide internal quality control to identify images that were of too poor quality for reliable interpretation, and to reassure the user that the images provided were acceptable.

Several doctors raised the matter of point-of-care ultrasound (POCUS). Several of them already have some experience with POCUS and use it in their practice on a day-to-day basis. They expressed an interest in a tool that would guide them to capture good renal US images, as well as assist with basic interpretation.

Current use of digital tools and technology

All doctors interviewed indicated that they have ready access to the internet; some have in-hospital Wi-Fi but many use their own data. Most indicated that they would consider it acceptable to use their own data to use a tool such as this, as they already use their data to access other applications for work-related purposes. All of them stated that they mostly use mobile devices and have limited access to a regular desktop, and so any application should be mobile-friendly.

Every doctor we interviewed was familiar with the tools EMGuidance (a medical reference application developed in SA) and Vula, an application which facilitates structured digital referrals. Younger interviewees, in particular, were using these tools in practice. Radiologists we interviewed also used websites such as STATdx and myelination MRI atlas as reference guides to assist their imaging interpretation.[22,23] One senior specialist noted that all clinicians must possess a smartphone because that is how they are paged, making it essentially an unspoken requirement for practice. Doctors reported using messaging tools like WhatsApp to communicate with each other. These tools and the ubiquity of mobile phones were pointed to as a lifeline for younger, less experienced staff who are fielding a wide array of medical issues, particularly before they enter a specialist training programme. This landscape of technology represents a fantastic opportunity to integrate tools where already tech-savvy clinical users access them both in centralised institutions via PACS and in a decentralised fashion via cell phone applications.

Clinician comfort with artificial intelligence

We found the doctors interviewed to be quite familiar with AI algorithms as a concept within and outside the medical sphere. Even those who expressed doubts about the ability to acquire acceptable quality US images from referral centres embraced the effort but noted the need for validation of any tool within the context of its use. That is, no doctors felt comfortable using the HN tool originally described as an off-the-shelf algorithm, but all felt that they would be comfortable using a similar tool that was adapted and validated with South African data, and with permission from their own or referring institution's specialists, or the provincial Health Department in some cases. One interviewee mentioned that they would like to know how users could provide feedback to the algorithm (that is, inform the AI tool that it had made an error).

Numerous respondents felt that a tool such as this would provide them with 'backup' in terms of their own requests and decisions. Doctors at a primary level expressed a belief that their consultations requests might be taken more seriously if accompanied by what would perhaps be perceived as a more objective report from the AI tool, and those at specialist facilities thought that perhaps referring doctors would better accept their feedback, also if accompanied by a report from a validated AI tool.

Conclusion

An AI tool designed to assist with the diagnosis and management of HN would potentially be well received and utilised in SA. The precedent for digital referral pathways has already been set by units and institutions making use of WhatsApp and Vula to facilitate referrals. There is a need to increase awareness of renal disease in children amongst treating clinicians, but, in addition to this, these clinicians would benefit from tools that help them to create structured referrals and prognosticate on their patients. A tool that is able to offer some interpretation of paediatric US images as well as receive inputs on other clinical features including patient history, height, weight, serum creatinine and urine microbiology would potentially assist with this. Clinicians interviewed felt that such a tool would not only help them to better motivate for the transfer of patients requiring tertiary care but would also save some patients from making a long journey to a referral hospital for no reason. We believe that a tool such as this may have a role to play in improving renal outcomes in the South African population.

Improving US acquisition and interpretation also represents a major opportunity for AI models to improve care. Ultrasound-assisted AI technology could have impacts far beyond HN clinical management or renal US alone and could be applied to many other US types and disorders. US is one of the cheapest, least invasive imaging modalities. Tools that make US easier to perform and interpret will empower more junior clinicians, sonographers and radiologists as well as those with less exposure to paediatric patients, in turn, also facilitating communication between these groups.

Study limitations

The current work represents clinicians working in seven of the nine provinces in SA and only one primary care medical officer was interviewed for this study. We believe this limitation is mitigated by the fact that most clinicians interviewed had prior experience working in other provinces as well as interacting with patients and clinicians in other provinces through referrals. In addition, all clinicians above the medical officer level (registrar/fellow, specialist) had completed a year or more of primary care service and spoke to their experiences in this environment.

Another limitation of this work is the focus on clinicians and the clinical environment in isolation of AI and digital tool development in SA. That is, our team's AI expertise is currently solely located in North America, thus leading to the potential for "offshoring" of future computational work. Therefore, future work will engage with data scientists and technical experts within SA to build toward more sustainable tool development, deployment and maintenance.

In conclusion, medical practitioners are generally enthusiastic about the potential utility of an AI tool to assist with the diagnosis and management of children with HN

in SA, provided the tool was validated for the SA context. Potential limitations to the use of such a tool include the quality of available imaging and getting broad-based 'buy-in' on both the referring and receiving end of the patient pathway. The further development, validation and clinical integration of these tools into new and existing applications have the potential to measurably improve outcomes for at-risk patients.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethical approval for this study was obtained from the Human Research Ethics Committee at the University of the Witwatersrand (Protocol number: M211119) and the Research Ethics Board at the Hospital for Sick Children (REB number: 1000079335).

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Appendix A

Interview to be conducted via Zoom – pilot topic areas and questions

Clinical setting

Here we wanted to unpack the experience of clinicians who see these patients. First, we demonstrate our AI tool that was built to assist with HN clinical management.

This will help us gauge how they view HN as an issue, given their clinical setting, and thus how as well as whether they would use a tool of this kind:

- Is HN front-of-mind here?
- If HN is considered a problem, how is it being thought about?
- Do you currently use digital tools for your clinic?
- If you had infinite choice, what would be the ideal AI/digital tool for your clinic?
    - Tools that assist with diagnosis? Therapy options? Follow-up/referral pathways?
- What reservations would you have with using an AI/digital tool?
    - Do you see any issues with using a tool that interprets ultrasound to stratify high- vs low-risk HN patients? (E.g., from the perspective of your patients and practice or in terms of clinical loads for yourself or others.)
    - Do you think you would ever consider using one of these tools, not necessarily just for hydro, but for other conditions also?
        - If not, why not, what more would you want to know, what reservations do you have?
        - Do you feel that the tool would be very unlikely to contribute to your process?
        - Do you have moral or ethical reservations regarding artificial intelligence? (E.g., privacy, data ownership, model getting it wrong, etc.)
        - Do you think it would be too complicated to use or add too much extra time to the clinical contact session, etc.?
    - Do you think that even if you do not use the tool for your decision-making process, that it would be useful for clinicians who need to refer to you?
    - Do you think it would be practical for district-level practitioners to use a tool like this to guide them regarding next steps once presented with a patient with hydronephrosis?
    - What would you like to see in a tool like this?

Technology access

Here we wanted to better understand the current technological capabilities and practices in different clinical settings. This will help us to better understand (1) how an AI tool would ideally be delivered (i.e., phone/internet/etc.) and (2) what kinds of upgrades may work to better enable these tools (i.e., ultrasound machine/probe, data storage):

- Is Wi-Fi reliably available?
- Are there smart phones? What kind(s)?
- Desktop/laptop computers? What kind(s)?
- Data storage? What kind(s)?
- Ultrasound machines available? What kind(s)?
    - What kind of ultrasound images (paper image, computer file, etc.) are you getting and how are they stored?

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PAEDIATRIC SURGERY

Analysis of mortality risk factors for newborns with Bochdalek diaphragmatic hernia – a 10-year single-centre experience

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Background and objectives: In this study, we aimed to investigate the risk factors for mortality in Bochdalek diaphragmatic hernia (BDH) patients.
Methods: Patients who underwent BDH treatment in the Paediatric Surgery Clinic of Dicle University, Turkey, between January 2010 and January 2020 were evaluated retrospectively. We recorded each patient's sex, hernia side, maternal and gestational ages, delivery type, birth weight, transfer information, blood gas values, Apgar scores at 1 and 5 minutes, pulmonary hypertension (PHT), time of death, surgical information, and any accompanying anomalies.
Results: There were 74 patients in our study, including 46 (62%) males and 28 (38%) females. Maternal age, birth week and weight, and mode of delivery did not differ significantly between the patients who are alive or have died. Mortality was higher in patients with abnormal preoperative blood gas values, 1 and 5 minutes Apgar scores below 7, and high blood lactate levels ($p < 0.05$). In total, 37 (50%) of 74 of our patients died. Concerning the time of death, 41% of the patients died within the first 24 hours, and 41% died between 24 hours and 7 days. Thirty-five patients died within the first month, and two died within 4–6 months due to additional problems. Over the long-term (10 year) follow-up of our surviving patients, three (8%) of 37 had recurrent respiratory infections, three (8%) had neurological problems, and two (5%) had gastro-oesophageal reflux. Present mortality also as a percentage (50%).
Conclusion: In our study, abnormal blood gas values, a low Apgar score, PHT, major and severe heart anomalies, and transfer from an external centre in an unstable state were statistically significant risk factors for mortality. The blood lactate level was significantly higher in patients who died. This can be considered a novel risk factor for mortality in patients with BDH.
Keywords: mortality, newborns, Bochdalek, diaphragmatic hernia

Introduction

Congenital diaphragmatic hernia is a congenital anomaly seen in 1 in every 3 000 live births, characterised by herniation of intra-abdominal organs to the thorax. It is one of the most common major congenital anomalies worldwide. More than 95% of such hernias are Bochdalek diaphragmatic hernias (BDHs). Posterolateral left-sided localisation is seen most commonly (six times more often than on the right side). If lung function is affected, respiratory problems may cause symptoms such as cyanosis in the first few hours after birth.[1-4] Despite advances in neonatal intensive care, mortality and morbidity rates are still high; mortality rates of 30–40% and 30–62% in developed and developing countries have been reported, respectively. In addition, the mortality rate can be as high as 82% due to various factors and comorbidities, with pulmonary hypertension (PHT) and hypoplasia being the most important factors in terms of both postnatal mortality and morbidity.[5-8]

In total, 40–50% of patients with BDH also have abnormalities of the cardiovascular, central nervous, genitourinary, or gastrointestinal system, and/or of chromosomes. Therefore, a multidisciplinary approach is required for the treatment of these patients.[9,10]

Many prognostic factors, such as low birth weight, Apgar score, prematurity, liver herniation, and blood gas values, have been investigated in patients with BDH, where mortality and morbidity vary depending on factors such as overall health status, region, and the experience of the centre.[11]

Although there have been important developments in intensive care, ventilator support, extracorporeal membrane oxygenation (ECMO) and PHT treatment in recent years, patients with BDH continue to present a serious challenge to clinicians. Improved treatment conditions have gradually increased survival; however, morbidity also increases with the long-term survival of these patients, so their management is becoming increasingly important. Determining the prognostic factors is important to reduce mortality.[11]

Over the long term, patients may experience problems such as chronic lung disease, gastro-oesophageal reflux, growth and development retardation, hearing loss, and neurodevelopmental issues.[12,13]

Table I: NSQIP-P cardiac risk factor categories, criteria and examples[39]

Category	Criteria	Example
None	1. No pre-existing cardiac condition	NA
	2. No compromise of cardiac function requiring medication	
Minor	1. Cardiac condition with or without medications and maintenance	Atrial septal defect; small to moderate ventricular septal defect with no symptoms
	2. S/P repair of congenital heart defect with normal cardiovascular function and no medications	Repaired atrial septal defect, ventricular septal defect, or patent ductus arteriosus
Major	1. S/P repair of congenital heart defect with residual haemodynamic abnormality with or without medications	Tetralogy of Fallot; aortic valve disease with aortic stenosis or insufficiency based on echocardiographic gradient, all single ventricle patients (including stage 1 repair)
Severe	1. Uncorrected cyanotic heart disease	Hypoplastic left heart syndrome; hypoplastic right ventricle; mitral atresia; single ventricle without repair; truncus arteriosus
	2. Any pulmonary hypertension	
	3. Ventricular dysfunction requiring medications	

In this study, we aimed to investigate the risk factors for mortality in BDH patients.

Materials and methods

Patients who underwent BDH treatment in the Paediatric Surgery Clinic of Dicle University, Turkey, between January 2010 and January 2020 were evaluated retrospectively. Our hospital is a high-volume university hospital in the southeast Anatolia region in Turkey.

Study design and patient groups

This study included newborn patients (< 1 month old) born in, or referred to, our hospital and diagnosed with BDH.

Any patient with signs of barotrauma whose pO_2 and pH could not be increased above 50 mmHg and 7.15 respectively, despite the application of conventional methods, was referred to an ECMO centre and excluded from the study.

Data were obtained retrospectively from patient files and computer records. Long-term follow-up sessions were held in the outpatient clinic, and patients who could not attend them were contacted by telephone. Cardiac risk was classified according to Table I.

We recorded each patient's sex, hernia side, maternal and gestational ages, delivery type, birth weight, transfer information, blood gas values, Apgar scores at 1 and 5 minutes, PHT, time of death, surgical information, and any accompanying anomalies. Long-term follow-up outcomes were also analysed; we divided the patients into two groups according to whether they died or survived and then compared these groups.

Management after hospitalisation

All deliveries of patients receiving a prenatal diagnosis at our hospital were planned in our centre. Patients born in other centres were transferred to our institution. All patients were intubated without mask ventilation in the early period; they were connected to a mechanical ventilator and a nasogastric tube was inserted. Temperature, glucose, and fluids were maintained at normal levels. Broad-spectrum antibiotics for gram-positive and -negative bacteria were started prophylactically. For all infants, mechanical ventilation was started at a high respiratory rate, and intermittent or continuous ventilation was then used according to the clinical needs of the patient. If intermittent or continuous ventilation was required, we used high-frequency oscillating ventilation. All infants with BDH were initially sedated with continuous intravenous midazolam. An umbilical catheter was preferred and a central venous line was installed. Metabolic acidosis was corrected using intravenous fluid and respiratory acidosis was corrected by adjusting the mechanical ventilator settings. We aimed for saturation > 85%. If severe PHT was present, we used intravenous sildenafil. Echocardiography and a posterior anterior chest radiograph were performed in every patient, and if these identified functional or structural heart disease, treatment was performed. Genitourinary examination and karyotype analysis were also performed.

Patients were considered stable when blood test results were normal for their gestational age (pH > 7.45, pCO_2 < 60 mmHg, pO_2 > 45 mmHg) and the preductal saturation level was 85–95%. Surgery was planned for patients with serum lactate < 3 mmol/L and urine output > 2 ml/kg/h.

Enteral nutrition was started on the first day after surgery and gradually increased if tolerated. Most patients underwent open surgery; before closing the diaphragm, we inserted a tube into the pleural space through the abdominal wall. Feeding via an orogastric tube was started on the first postoperative day. After 2–3 days, the tube in the pleural cavity was removed. The patients were followed for a long time (mean follow-up = 5 years; range 1–10 years).

Statistical analyses

We performed statistical analysis using SPSS software (version 21.0; SPSS Inc., Chicago, IL, USA). The Shapiro–Wilk test was used to determine whether the data had a normal distribution. Continuous, normally distributed variables were compared using student's t-test, while categorical variables were compared using the chi-square test. Non-parametric tests were used when the data were not normally distributed. Binary logistic regression method was used to analyse the relationship between the variables. We used receiver operating characteristic analysis to evaluate our predictive model. We considered a p-value < 0.05 statistically significant.

Results

There were 74 patients in our study, 46 (62%) males and 28 (38%) females. Among the patients, 59% had a maternal age > 25 years, 72% were mature, 70% were born by caesarean section, and 77% had a birth weight > 2 500 g. In

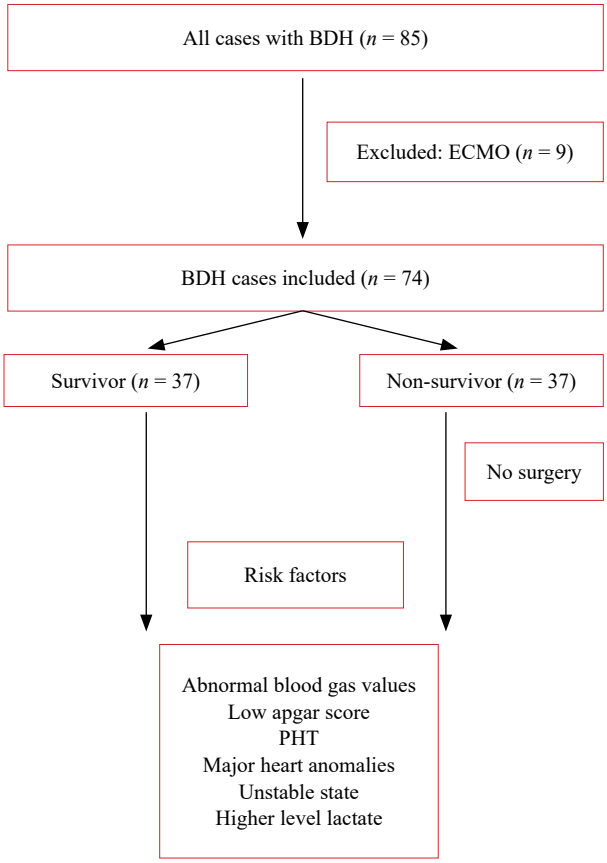

Figure 1: Analysis chart of mortality risk factors

total, 64% and 41% of the outborn and inborn patients were unstable, respectively. Maternal age, birth week and weight, and mode of delivery did not differ significantly between the living and deceased patients. Prenatal diagnosis was 74%, 62% of this was in survivors and 68% in non-survivors.

Mortality was higher in patients with abnormal preoperative blood gas values, 1- and 5-minute Apgar scores below 7 and high blood lactate levels ($p < 0.01$ or 0.05); patients with stable preoperative blood gas values had a lower mortality rate ($p < 0.05$). The mean lactate level was 3 mmol/L in alive patients and 8 mmol/L in deceased patients (Figure 1).

The most common type of accompanying anomaly was cardiac; this was seen in 37 (50%) patients who had a significantly higher mortality rate ($p < 0.01$). In total, 10 of 74 patients were unstable at presentation. Nine patients died before echocardiography could be performed; it was completed in 50 of the remaining patients, among whom major heart anomalies were the most common issue (43%). Nineteen per cent of severe anomalies and 28% of minor anomalies died ($p < 0,01$). Twelve patients had PHT; sildenafil (a phosphodiesterase inhibitor) was used in the five patients with a severe form of PHT, and PHT significantly increased mortality ($p < 0.05$). Eighty per cent of the unstable patients from the outborn died ($p < 0.05$).

Surgery was performed in 88% of all patients but could not be performed in 12%. Surgery was performed during the first 48 hours in 80% of the patients operated on. Laparotomy was the surgical method used in 97% of the surgical patients. The posterior rim of the diaphragm was absent in 20% of the surgical patients, whose diaphragms moved freely. In total, 82% of the diaphragmatic defects

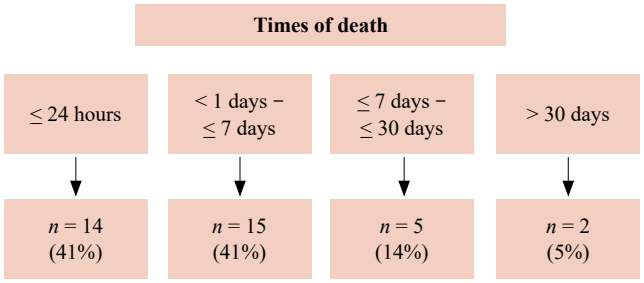

Figure 2: Times of death

were treated via primary repair, with the remaining 18% repaired with a mesh. The most frequently herniated organs were the small intestine (78%) and colon (72%), and the least frequently herniated was the kidney (6%). In total, 40% of surgical patients underwent the Ladd procedure for malrotation. Postoperative complications were seen in four patients, two of whom had recurrence; the other two had wound infections. When we compared the deceased and surviving patients, no difference was found in terms of surgical time, type of surgery, complications, or the organs herniated. Over the long-term (10 year) follow-up of our surviving patients, three (8%) of 37 had recurrent respiratory infections, three (8%) had neurological problems, and two (5%) had gastro-oesophageal reflux.

In total, 37 (50%) of 74 of our patients died. Concerning the time of death, 41% of the patients died within the first 24 hours, and 41% died between 24 hours and 7 days. Thirty-five patients died within the first month, and two died within 4–6 months due to additional problems (Figure 2). The mean follow-up period was 5 years (range 1–10 years). Detailed results of the analyses of our patients are listed in Tables II and III.

Discussion

Evaluation of the treatment results of BDH is very difficult due to differences among clinics in terms of treatment strategy, patient risk, ventilation mode, ventilation strategy and pharmacological agents. Therefore, many factors should be considered in the prognosis of a newborn with BDH. Moreover, it is important to analyse both short- and long-term results to keep high-risk patients alive and to evaluate the side effects of the treatments applied.

In the literature, the male-to-female ratio for BDH is approximately 1.5–2:1;[14] in our study, it was 1.6:1.

BDH is more common on the left side; the left-right ratio varies between 4:1 and 9:1, and mortality is higher for cases of right-sided hernia.[14] In our study, BDH occurred approximately six times more frequently on the left than right side, and the mortality rate was 79% and 21%, respectively which was consistent with the literature.

The mortality rate has been reported as 75% in premature patients, but no significant relationship between mortality and the week of birth has been reported in the literature.[15] Also, although low birth weight has been reported as an indicator of poor prognosis, some studies did not find this.[16] In our study, we found that the mortality rate was 75% (9/12) in premature patients, but no statistically significant difference from the other births was found. We believe that this was due to the low proportion of premature patients (12/43; 28%) in our study. Thus, a large case series is

Table II: Patient characteristics in the two groups

		Overall *n* = 74	Survivors (Mean ± SD or %) *n* = 37 (50%)	Non-survivors (Mean ± SD or %) *n* = 37 (50%)	*p*	*p**
Sex	Male	46 (62%)	25/37 (68%)	21/37 (57%)	$p > 0.05$	$p > 0.05$
	Female	28 (38%)	12/37 (32%)	16/37 (43%)		
Side of lesion (right vs left)	Left	56/65 (86%)	34/37 (92%)	22/28 (79%)	$p > 0.05$	$p > 0.05$
	Right	9/65 (14%)	3/37 (8%)	6/28 (21%)		
Maternal age	> 25 years	17/29 (59%)	5/9 (56%)	12/20 (60%)	$p > 0.05$	$p > 0.05$
	< 25 years	12/29 (41%)	4/9 (44%)	8/20 (40%)		
Gestational age (GA) in weeks	> 37	31/43 (72%)	11/14 (79%)	20/29 (69%)	$p > 0.05$	$p > 0.05$
	< 37	12/43 (28%)	3/14 (21%)	9/29 (31%)		
Delivery mode	Caesarean	28/40 (70%)	12/16 (75%)	16/24 (67%)	$p > 0.05$	$p > 0.05$
	Vaginal	12/40 (30%)	4/16 (25%)	8/24 (33%)		
Birth weight (BW)	> 2 500	46/60 (77%)	23/27 (85%)	23/33 (70%)		
	2 000–2 500	8/60 (13%)	3/27 (11%)	5/33 (15%)	$p > 0.05$	$p > 0.05$
	< 2 000	6/60 (10%)	1/27 (4%)	5/33 (15%)		
Patient situation						
Inborn 49/74 (66%)	Stable	29/49 (59%)	17/22 (77%)	12/27 (44%)	$p < 0.05$	
	Unstable	20/49 (41%)	5/22 (23%)	15/27 (56%)		$p < 0.05$
Outborn 25/74 (34%)	Stable	9/25 (36%)	6/10 (60%)	3/16 (19%)	$p < 0.05$	
	Unstable	16/25 (64%)	4/10 (40%)	12/16 (81%)		
Preoperative blood gas values	pH		7.4 ± 0.1 (7–7.5)	7.1 ± 0.3 (6.5–7.4)	$p < 0.01$	
	pO_2		79.6 ± 23 (60–110)	51.1 ± 23 (30–90)	$p < 0.01$	
	HCO_3		21 ± 2.5 (19–29)	17 ± 5.4 (10–28)	$p < 0.01$	$p < 0.05$
	CO_2		35.9 ± 8 (22–50)	65 ± 42 (33–130)	$p < 0.01$	
	Lactate		3 ± 1.7 (1.8–4.5)	8 ± 7 (2–24)	$p < 0.01$	
Apgar score	Apgar at 1 min		6 ± 1 (5–8)	3 ± 1.7 (1–6)	$p < 0.01$	$p < 0.05$
	Apgar at 5 min		8 ± 1 (7–10)	5.7 ± 1.4 (3–8)		
Prenatal diagnosis		55/74 (74%)	23/74 (62%)	25/74 (68%)	$p > 0.05$	$p > 0.05$
Total length of stay		days	14 ± 7.8 (4–33)	5 ± 8.2 (0.04–40)	$p < 0.01$	
Time surgery		minutes	105 (60–240)	111 (60–180)	$p > 0.05$	$p > 0.05$
Associated anomaly	Heart	37/74 (50%)	22/37 (59%)	15/37 (41%)		
	SSS	9/74 (12%)	5/37 (14%)	4/37 (11%)		
	Urogenital	4/74 (5%)	2/37 (5%)	2/37 (5%)		
	Gastrointestinal	2/74 (3%)	1/37 (3%)	1/37 (3%)	$p > 0.05$	$p > 0.05$
	Chromosomal	1/74 (1%)		1/37 (3%)		
	Endocrine	1/74 (1%)	1/37 (3%)			
Heart pathology	Normal	10/50 (20%)	8/29 (28%)	2/21 (10%)		
	Minor	24/50 (48%)	18/29 (62%)	6/21 (28%)		
	Major	12/50 (24%)	3/29 (10%)	9/21 (43%)	$p < 0.01$	$p < 0.05$
	Severe	4/50 (8%)	0	4/21 (19%)		
Pulmonary hypertension (pre-repair)	Yes	12/74 (16%)	2/37 (5%)	10/37 (27%)	$p < 0.05$	$p < 0.05$
Pulmonary vasodilators for PH	Sildenafil	5	1 (20%)	4 (80%)	$p > 0.05$	$p > 0.05$
Long-term complications	Recurrent respiratory infections		3 (8%)			
	Neurological problems		3 (8%)			
	Gastro-oesophageal reflux disease		2 (5%)			
	Recurrence of CDH		1 (3%)			
	Small bowel obstruction		1 (3%)			
	Pulmonary abnormalities		1 (3%)			
	Recurrence of CDH		1 (3%)			
	Scoliosis		1 (3%)			
	Failure to thrive		1 (3%)			

*p** – logistic regression analyses

Table III: Operative findings in the two groups

		Overall $n = 74$	Survivors (Mean ± SD or %) $n = 37$ (50%)	Non-survivors (Mean ± SD or %) $n = 37$ (50%)	p
Surgical management	Surgery	65 (88%)	37 (100%)	28 (76%)	$p > 0.05$
	No surgery	9 (12%)	0	9 (24%)	
Time of operative repair	Surgery < 48	52 (80%)	26 (70%)	26 (93%)	$p > 0.05$
	Surgery > 48	13 (20%)	11 (30%)	2 (7%)	
Type of surgery	Laparotomy	63 (97%)	35 (95%)	28 (100%)	$p > 0.05$
	Thoracoscopy	1 (1.5%)	1 (2.5%)		
	Laparoscopy	1 (1.5%)	1 (2.5%)		
Posterior rim of the diaphragm	No	13/65 (20%)	8/37 (22%)	5/28 (18%)	$p > 0.05$
Repair type	Primary	53 (82%)	28/37 (76%)	25 (89%)	$p > 0.05$
	Patch/Flap	12 (18%)	9/37 (24%)	3 (11%)	
Herniated organs	Small bowel	51/65 (78%)	27/37 (73%)	24/28 (86%)	$p > 0.05$
	Colon	47/65 (72%)	23/37 (62%)	24/28 (86%)	
	Stomach	42/65 (65%)	20/37 (54%)	22/28 (79%)	
	Spleen	37/65 (57%)	20/37 (54%)	17/28 (61%)	
	Liver herniation left CDH	14/65 (22%)	6/37 (16%)	8/28 (29%)	
	Liver herniation right CDH	5/65 (8%)	2/37 (5%)	3/28 (11%)	
	Kidney	4/65 (6%)	2/37 (5%)	2/28 (7%)	
Other operation	Ladd procedure	26/65 (40%)	14/37 (38%)	12/28 (43%)	$p > 0.05$
	Appendectomy	8/65 (12%)	6/37 (16%)	2/28 (7%)	
	Abdominoplasty	1 (1.5%)	1 (3%)	0	
Operative repair	Primary surgery	63/65 (97%)	36/37 (97%)	27/28 (96%)	$p > 0.05$
	Seconder surgery	2/65 (3%)	1/37 (3%)	1/28 (4%)	
	Surgical complication	4/65 (6%)	2/37 (5%)	2/28 (7%)	
	Resurgery	2/65 (3%)	1/37 (3%)	1/28 (4%)	
Postoperative complications	Recurrence	2/65 (3%)	1/37 (3%)	1/28 (4%)	$p > 0.05$
	Wound infection	2/65 (3%)	1/37 (3%)	1/28 (4%)	
Chest drain inserted	Yes	65	37/37 (100%)	28/28 (100%)	$p > 0.05$

needed to identify the relationship between prematurity and mortality.

Hidaka et al. and Lazar et al.[17,18] reported that mortality was significantly higher for BDH infants with a birth weight under 2 500 g, although Takahashi et al.[11] found no relationship between birth weight and mortality. In addition, the latter study found no relationship between mode of delivery and mortality.[11,17,18] In our study, we found no significant relationship between birth weight and mortality. We believe that this was due to the low proportion of patients with low birth weight (23%).

It is important for mothers to give birth in the centre where the surgery will be performed, because patients with BDH born in external centres can become unstable during transfer. In studies by Wynn et al. and Teo et al.,[19,20] 37% and 2%, respectively, of the patients were born in an external centre. In our study, 34% of patients were referred from external centres, of whom 40% survived and 60% died. Some of the patients from external centres became unstable due to incorrect intubation, increased pressure in the lungs due to gastrointestinal problems, insufficient sedation and inexperienced transfer personnel. Some of them died before surgery. Therefore, we believe that such patients should be delivered in experienced centres where the surgery will be performed.

The relationship between blood gas analysis and mortality has been investigated as a prognostic factor by Boix-Ochoa et al.[21-25] In early studies, significant differences were found in pH and pCO_2 between surviving and deceased patients; low pCO_2 and high pO_2 values are considered good prognostic indicators. Also, a high pCO_2 despite mechanical ventilation has been reported as an indicator of poor prognosis. Gentili et al.[22] reported that the blood gas values (pH > 7.35 and pCO_2 < 55 mmHg) of patients with BDH can be used to predict the prognosis. Haricharan et al.[23] reported that pCO_2 > 60 mmHg in the early period was associated with high mortality, and Hoffman et al.[25] similarly reported that survival was 27% when $paCO_2$ < 60 mmHg in the first blood gases; all patients with pCO_2 > 70 mmHg died.[10,21,23-25] In our study, mortality increased significantly in patients with $paCO_2$ levels > 60 mmHg before surgery ($p < 0.01$). Also, the prognosis of patients with pH > 7.35 was better ($p < 0.01$). Based on these results, low pH and high pCO_2 are risk factors for mortality.

In our study, we also found that a high blood lactate level increased mortality significantly ($p < 0.01$). The literature contains only a limited number of studies reporting the lactate levels of patients with BDH.[21,23-25] Thus, we have made a novel contribution to the literature on risk factors in patients with BDH.

Studies have shown that patients with low Apgar scores at 1 and 5 minutes have higher mortality.[26] Similarly, we observed a significantly higher mortality rate ($p < 0.01$) in such patients.

Additional abnormalities in these patients may affect the prognosis. In the literature, the rate of additional abnormalities varies between 10% and 60%; in particular, the rate of cardiac malformations ranges between 10% and 20%. In the study of Takahashi et al.,[11] 32% of patients had serious heart problems, the most common being ventricular septal defect.[10,11,27] In our study, cardiac malformation was the most common cardiac problem, accounting for 48% of the cases with minor cardiac anomalies. In addition, major cardiac problems were detected in 24% of our patients, of whom 9/12 (75%) died, and all 8% of patients with severe heart problems died. Therefore, mortality increased with the severity of cardiac anomaly.

In newborns with BDH, PHT is a common problem; it results from factors such as hypoxia, hypercarbia, acidosis and hypothermia that persist in the foetal circulation system, and is the most common cause of mortality in BDH patients. In a study by Sekhon et al.,[28] PHT was detected in 100% of the patients who died. In a study by Bojanić et al.,[10] PHT was detected in 86% of the patients who died.[28,29] In our study, similar to the literature, 10/12 (83%) of patients with PHT died. Sildenafil has been used to treat PHT patients.[30] In our study, we used sildenafil in the 40% of patients with PHT, of whom 20% survived.

In the modern surgical management of CDH, semi-elective surgery is performed after initial stabilisation.[30] In our study, 88% of the patients underwent surgery; the other 12% died before surgery could be performed because they could not be stabilised.

Treatment of BDH occurs in response to a physiological emergency, not a surgical one; the patient should be stabilised before surgery, which can take several days. Surgery was performed at 24–48 and 3–12 days in the studies of McHoney et al. and Terui et al., respectively.[31,32] In our study, the average time to surgery was 3.4 days among patients who died and 5.2 days among those who survived. In a study by Rozmiarek et al.,[33] there was no significant difference in mortality between patients who underwent surgery before versus after the first 48 hours.[33] Therefore, we recommend performing surgery when the patient is stable, rather than within the first 48 hours.

Patch use is required in approximately half of all BDH patients, with some studies reporting its use in 15–84% of cases.[30-35] In our study, we preferred primary repair, and so used patches in only 18% of our patients; this is a low rate compared to the literature. Accordingly, our recurrence rate was not high. In patients without a posterior rim, we anchored sutures around the posterior ribs; this method appears to reduce both the use of patches and the recurrence rate.

Herniated organs as a prognostic criterion in BDH has become controversial; another possible criterion is the formation of the liver and stomach in the thorax.[14] In the study of Gentili et al., liver and gastric hernias were found in 60% and 50% of patients, respectively.[22] In our study, the most common hernia site was the small intestine (78%), followed by the colon (72%) and stomach (65%). With herniation of these organs, patients may experience respiratory distress when breathing with a mask. In total, 57% of our patients with left diaphragmatic hernias died. In all five patients with right diaphragmatic hernia, the liver was herniated; 60% of these patients died, but statistical significance was not attained because the number of patients was small. Liver herniation on the right side appeared to increase mortality, as in the literature.

Additional surgical approaches vary among clinics; while some do not recommend correcting malrotation, others suggest complete or partial correction during the first surgery. Surgeons who recommend surgical correction argue that it reduces the future risk of volvulus. The malrotation rate was reported to be 42–60% in a study by Heiwegen et al.[35] In our study, malrotation was present in 40% of the 65 patients who underwent surgery and was corrected in all patients; we performed an appendectomy in 13% of these patients. We did not observe volvulus in any of the patients over the long-term follow-up.

In one study, surgical complications were found in 14% and 20% of the surviving and deceased patients, respectively, with repeated surgery needed in 9% and 20% of these cases, respectively.[7] In our study, the overall rate of surgical complications was low (6%). One patient relapsed during the first month, and a second patient relapsed at 4 months; a patch was used for both. In addition, there was one case of wound infection in each group.

Although different clinics report varying outcomes for BDH patients, the mortality rate of this disease is still high, at 25–83%, with a mean of 60%.[5,6,20,37-39] Some newborns die before they can even be operated upon, with Geary et al.[38] reporting that 11 out of 34 newborns (32%) died in this way due to additional problems and PHT.

In our study, the mortality rate was 50% without ECMO. One of the factors that increases mortality according to some studies is treatment in a regional hospital, so patients with additional problems were referred to our institution. Some of our patients were transferred from an external centre in an unstable state and died within a few hours. Therefore, BDH patients should be delivered in a reference hospital or only transferred once their condition has improved. In one study, nine infants died during the first 24 hours (9.5%), 11 died at 1−28 days (11.6%), and one died after day 28 (1.1%). In total, 21 infants died within 1 year.[26] In our study, 41% of the patients died within the first 24 hours, 41% within 1–7 days, 14% within 7–30 days, and 5% after 30 days.

In the literature, many factors such as PHT, postpartum instability, cardiac and other anomalies, sepsis, low birth weight, and prematurity have been reported as causes of mortality in BDH patients.[20,30,34] In our study, one patient each died from sepsis and cardiac anomaly, sepsis and renal failure, and cardiopulmonary instability. The other patients died due to multiple additional problems and instability.

Health problems that occur later in life in BDH patients may require treatment. Although some problems regress spontaneously over time, or with medical treatment, other pathologies are difficult to detect at a young age but manifest at an advanced age and need treatment. Gastro-oesophageal reflux, PHT, and respiratory and neurological problems tend to regress with age. On the other hand, problems of the musculoskeletal system, such as chest deformities and scoliosis, are expected to worsen with age. The incidence of these problems continues to increase rapidly even with the proliferation of treatment approaches.[20,30,34] In our study, two patients (6%) had long-term gastro-oesophageal reflux

problems; we treated these patients medically. Intestinal obstruction due to adhesion occurred in one patient at 6 months postoperatively and was corrected surgically. Growth and development retardation were detected over the long-term follow-up in one patient in the 9% percentile.

Although ECMO increases the survival rate of severe BDH patients, it can have chronic consequences such as growth retardation and neurodevelopmental or hearing problems, which affect survivors' aerobic exercise capacity.[10] One limitation of our study was that this group of patients could not be included, because the study was not performed in our ECMO centre.

Conclusion

In our study, abnormal blood gas values, a low Apgar score, PHT, major and severe heart anomalies, and transfer from an external centre in an unstable state were statistically significant risk factors for mortality ($p < 0.05$). Gender, maternal age, mode of delivery, birth weight and type of operation did not affect mortality. The blood lactate level was significantly higher in patients who died ($p < 0.01$); this can be considered a novel risk factor for mortality in patients with BDH.

Conflict of interest

The authors report no conflict of interest.

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Ethical approval

The study was approved by the Ethics Committee of Dicle University (June 30, 2021; approval number: 353).

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PAEDIATRIC SURGERY

Presentation and management of choledochal cyst in children – personal experience and retrospective analysis of over 30 years from India

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Background: To study the varied presentations of paediatric choledochal cysts and to analyse the postoperative outcomes of two widely used anastomotic techniques – hepatico-duodenostomy (HD) and hepatico-jejunostomy (HJ). Such comparison would help establish the superiority of one procedure over the other, since the choice of bilioenteric bypass is debatable in cases of paediatric choledochal cyst.
Methods: Records spanning 30 years showed various presentations in 98 cases of choledochal cyst who reported with jaundice, abdominal pain, etc. Excision of cyst followed by bilioenteric bypass was done after necessary investigations and preoperative optimisation. Long- and short-term outcomes of HD and HJ were compared using statistical techniques. Jejunal graft inter-positioning hepatico-duodenostomy (JGIHD) was also performed in a small group of patients, the outcome of which was not included in the statistical analysis.
Results: Type I choledochal cyst was the most common pathology (n = 87; 88.7%) and recurrent abdominal pain was the commonest associated symptom (n = 32; 32.6%). Of 98 patients, one died before operative procedures could be performed. Among the rest, all 97 patients underwent cyst excision, followed by HD (n = 68; 70.1%) or HJ (n = 23; 23.7%) or JGIHD (n = 6; 6.2%). No significant difference in risk of immediate and delayed complications was observed between HD and HJ ($p > 0.05$). However, mean duration of hospital stay was significantly lower for HD ($p = 0.029$).
Conclusion: HD being technically easier, less complicated and less time consuming than HJ, should be preferred over the later.
Keywords: choledochal cyst, children, bilioenteric bypass, hepatico-duodenostomy, hepatico-jejunostomy

Introduction

Choledochal cyst is a congenital anomaly of the bile duct, more commonly found in the paediatric age group.[1] It is more prevalent in the Asian population, and the majority of the cases are found in Japan.[2] Though the treatment of choice for the cyst itself is standardised as complete excision of cyst, the type of bilioenteric bypass that is done is still not standardised.[3] There are a few studies which have compared the outcomes of different bilioenteric anastomosis techniques, yet there are no definitive guidelines available to date.[4-6] Other than the bilioenteric bypass procedure, there may be different patterns of presentation and complications related to the disease and treatments, which are to be dealt with in different ways.

The present study aimed to analyse the different patterns of presentation of choledochal cyst in paediatric patients, different treatment modalities for their management, different treatment modalities for complications related to the disease itself and complications related to the definitive treatments of the disease. The outcomes of hepatico-duodenostomy (HD) and hepatico-jejunostomy (HJ) in terms of postoperative morbidities (paralytic ileus, anastomotic leak, relaparotomy for biliary peritonitis, biliary gastritis during late follow-up, postoperative revision surgery for repeated cholangitis) and hospital stay were also compared with relevant statistical techniques.

The authors believe that the results of this study will ultimately help to establish standardised management procedures in paediatric cases of choledochal cyst. Such a study comparing the outcomes of two commonly used anastomotic procedures is rare from the Indian perspective, and the authors believe that it will enrich the existing knowledge and experience on the subject.

Methods

This retrospective study was conducted based on records spanning over a period of 30 years (February 1991–January 2020). Details of paediatric choledochal cyst cases who reported at a tertiary care centre of Kolkata during the study period were obtained and included in the study. Follow-up records were also analysed to study their overall experience.

Patients presented with choledochal cyst bearing different symptoms, such as abdominal lump, jaundice, pancreatitis, cholangitis and recurrent abdominal pain. All these patients were primarily evaluated clinically and then by necessary radiological and biochemical investigations, such as ultrasonography (USG) of the abdomen, magnetic resonance cholangiopancreatography (MRCP) and hepatobiliary iminodiacetic acid (HIDA) scan. MRCP was started routinely in the later phase after it became available in Eastern India, where we are located. HIDA scan was done in patients of early infancy to differentiate between cystic type of biliary atresia and choledochal cysts.[7] Acute complications related to choledochal cysts, such as pancreatitis, cholangitis and biliary peritonitis, were first treated, and 6–8 weeks after the patient stabilised, definitive surgical intervention was done.

After pre-anaesthetic optimisation, as part of definitive operative intervention, patients were subjected to either complete excision of cyst or mucosectomy (Lilly's procedure – it was performed in cases where extensive pericystic fibrosis was found and dissecting out the cyst as whole from the porta was found to be difficult) or excision of only the extrahepatic cyst for type IVA disease, as per Todani's classification of choledochal cysts.[8] This was followed by either Roux-en-Y hepatico-jejunostomy (RYHJ) or HD or jejunal graft inter-positioning hepatico-duodenostomy (JGIHD). In cases of type IVB choledochal cyst, HJ was the preferred option due to short stump. Postoperative clinical and biochemical (liver function test) follow-up records of all these patients were analysed. These records spanned a minimum period of 1 year to a maximum of 18 years (patients operated earlier in the study timeline had scope of longer follow-up). USG and endoscopy were done during follow-up in some cases when indicated (e.g., in delayed postoperative abdominal pain).

Short-term outcomes that were considered included immediate postoperative morbidities (paralytic ileus, anastomotic leak, relaparotomy for biliary peritonitis) and hospital stay. Long-term outcomes included delayed complications like biliary gastritis and cholangitis (associated with anastomotic stricture). For comparison, both short-term and long-term outcomes of HD and HJ were considered. These outcomes were compared statistically with the help of the chi-square test (or Fisher's exact test) or independent t-test, whichever was applicable. *P*-value < 0.05 was set as statistically significant.

The JGIHD was performed in the early phase of this study. However, eventually, as it was noticed that even after being a complicated and more time-consuming procedure, it did not serve any additional short-term benefit, it was not performed further. Hence the outcomes of JGIHD were noted here, but not included in the statistical analysis.

Results

In our setup, a total of 98 patients presented with choledochal cyst. The most commonly encountered choledochal cyst was type 1, found in 87 cases (88.7%). Others were type IVA (n = 1; 1.0%), type IVB (n = 7; 7.1%) and forme fruste disease or type VI choledochal cyst (n = 3; 3.1%).

Out of these patients, 28 (28.6%) were males and 70 (71.4%) were female. Of them, 32 patients (32.6%) had a history of recurrent attacks of abdominal pain. Some (n = 16; 16.3%) presented with jaundice. Only seven patients (7.1%) had abdominal pain as their chief complaint, and seven other patients (7.1%) presented with only abdominal swelling/fullness. Some (n = 16; 16.3%) presented with jaundice. Of all patients, nine (9.1%) presented with typical features of abdominal pain, swelling and jaundice, and four (4.1%) had associated gallstone disease.

All patients primarily underwent USG abdomen (100%). In 47 cases (47.9%) a MRCP and in three patients (3.1%) a HIDA scan was done.

Six patients (6.1%) with pancreatitis were also treated conservatively at first. Patients who presented with cholangitis with septicaemia (n = 3; 3.1%) were first treated with endoscopic retrograde cholangiopancreatography (ERCP) guided nasobiliary drainage and broad-spectrum antibiotics. Some (n = 4; 4.1%) patients presenting with features of acute pancreatitis were eventually found to have choledochal cyst. In both the cases of cholangitis and pancreatitis, definitive surgery was done 6–8 weeks after primary presentation, after the acute episode was controlled. The patients (4.1%; 4/98) who presented with perforative biliary peritonitis were primarily treated with drainage and thorough peritoneal toileting, and after 6–8 weeks, definitive surgery was done. One such patient, who primarily presented with biliary peritonitis and underwent laparotomy, died of severe sepsis on day 4 and hence was excluded from further analysis. All the remaining 97 patients with choledochal cyst underwent definitive surgery after necessary investigations and preoperative optimisation.

Out of the 97 patients who underwent definitive surgery, in 81 patients (83.5%) complete excision of cyst could be achieved; Lilly's procedure was done in 15 patients (15.5%). Excision of the extrahepatic cyst was done in the only case of type IVA cyst (1.0%).

The most commonly performed bilioenteric bypass procedure was HD (n = 68; 70.1%), followed by RYHJ (n = 23; 23.7%) and JGIHD (n = 6; 6.2%). In the six patients who underwent JGIHD, five (83.3%) had normal outcome without any major complications other than minor surgical site infection, one patient (16.7%) developed an anastomotic leak which was managed conservatively.

On following-up the 91 patients who either underwent HJ or HD, no significant difference in the risk of immediate and delayed complications was observed between the two groups. From the former group, two out of 23 patients (8.7%) and one patient (1.5%) from the HD group were found to experience prolonged paralytic ileus (p = 0.312). The number of anastomosis leaks was comparable in both groups – three (13%) in the HJ and five (7.4%) in the HD group (p = 0.649). Anastomotic leak was noted by the presence of bilious content in the abdominal drain after the third postoperative day.[9] The majority of such patients (5/8; 62.5%) with anastomotic leak without any features of peritonitis were successfully treated conservatively. These patients were kept nil per mouth with the functional drain, which was kept during primary surgery, and adequate supportive measures (fluid resuscitation, broad-spectrum antibiotic and ultrasound-guided drainage whenever needed). Some others (3/8; 37.5%) developed biliary peritonitis subsequently, where the drain output was reduced but the patients developed features of peritonitis. This group of patients needed re-operation – of them, two were from the HD group and one was from the HJ group (p = 0.999). No significant difference in risk of developing biliary gastritis was observed between the two groups (1/23; 4.3% in HJ

and 4/68; 5.9% in HD; $p = 0.999$). Recurrent episodes of cholangitis were seen in two patients (8.7%) in the HJ group, who ultimately developed anastomotic stricture and needed revision surgery (after 2 years in one patient and after 15 years in the other case); no one in the HD group developed cholangitis ($p = 0.124$). There was no mortality in either group. However, mean duration of hospital stay was significantly lower in the HD group (7.68 days; SD = 2.72) than in the HJ group (9.9 days; SD = 3.60; $p = 0.029$).

Discussion

Choledochal cyst is the commonest cystic disease of the bile duct which can lead to different clinical presentations.[2] Before going for surgical intervention, definitive radiological evaluation of the character/classification of the disease should be done as the procedure of choice varies in different types of choledochal cyst.[10]

Though it is rare, in 1–2% of cases, choledochal cyst patients may present with spontaneous rupture of the cyst followed by biliary peritonitis.[11,12] In our study, 4.1% had this complication during first presentation. Other presenting features which can lead to serious morbidities in children are cholangitis and pancreatitis.[11,13] Compared to existing literature, in our series of patients, cholangitis and pancreatitis were found in fewer patients.[2,13,14] For patients with acute cholangitis and peritonitis, it is always desirable to control the sepsis primarily by endoscopic intervention or surgical intervention; 6–8 weeks after the patient stabilises, definite surgical intervention is to be done. Patients with acute pancreatitis are also treated in the same manner.[10]

There are a handful of studies available at present which have compared the outcomes of HJ and HD. However, no such study from India was found.

The choice between these two anastomotic procedures (HD and HJ) is still a matter of debate. In their studies, Yeung et al., Todani et al. and Guzman et al. have shown that HD is not inferior to HJ.[4,5,15] Rather, HD has been shown to be better in some aspects, such as being less time consuming, more physiological and with less chance of postoperative ileus. On the other hand, in some studies, such as in Shimotakahara et al., Singham et al. and Machado et al., HJ has been preferred over HD for a few reasons, such as lesser risk of biliary gastritis, etc.[3,6,16]

In our experience, HD is less complicated and technically easier, as fewer anastomoses are required, and therefore it is arguably associated with lesser risk of anastomotic failure. In our study, cases of anastomosis leak and biliary peritonitis were not significantly different between both groups (HD vs HJ), the majority of which could be successfully treated conservatively.

Narayanan et al. showed in their meta-analysis of six retrospective studies that the average anastomotic leak is around 2–2.5% in both the HJ and the HD group. In our study, though the leak rate is a bit higher (13% and 7.4% in HJ and HD, respectively), the difference is not statistically significant. There had been no associated mortality because of the anastomotic leak. Most of these patients had been managed conservatively with a prophylactic drain placed during surgery and other supportive care, and this is also supported by the existing literature.[17,18]

Postoperative unusual abdominal pain with low-grade fever and unexplained leucocytosis must raise the suspicion of intraperitoneal bile collection. In such cases of patients with postoperative biliary peritonitis, the decision to open up the abdomen is vital, depending on the clinical and radiological features. If the bile remains sterile, it may take time to develop infective peritonitis, as it progresses in a subtle manner before development of gross peritonitis.[19]

In bilioenteric anastomosis, when the patient develops biliary peritonitis, during laparotomy, if the site of anastomosis leak is easily visible, then it can be repaired. Otherwise, thorough peritoneal toileting with adequate drain placement will suffice. Dissecting out an obscured anastomosis leakage at the anastomotic site with the intention to repair it is not needed, as bile being immunogenic and fibrogenic causes inflammation around the leaked anastomosis, eventually leading to fibrosis due to over-healing.[20] A minor amount of bile gets absorbed in the peritoneum.[10,21] This gradually seals off the leak and the distal main path, which remains patent, starts working after return of peristalsis.

In long-term follow-up a small proportion of patients might develop anastomotic stricture and cholangitis. A few studies have shown a higher number of stricture and cholangitis in the HJ group whereas a few other studies have shown the reverse.[3,22-25] In this study, we found two cases in the HJ group which developed anastomotic stricture and cholangitis and none among the HD group. However, it was not statistically significant.

According to existing literature, biliogastric regurgitation remains one of the major concerns in patients undergoing HD, which ultimately leads to malignancy.[23,26,27] However, the risk of development of malignancy is still a matter of debate.[28] In this study, we encountered no such findings. Over the period of 30 years, none of the patients developed cholangiocarcinoma. The total number of biliary gastritis, however, was higher in the HD group but was not statistically significant. In such cases, diagnosis was confirmed with upper GI endoscopy and successful resolution was achieved with proton-pump inhibitors and/or sucralfate. For the only case of biliary gastritis in the HJ group, barium meal x-ray showed distal kinking of the gut loop. The patient was given prokinetic first, but later needed laparoscopic adhesiolysis. In HD, biliary gastritis can be avoided with construction of the HD anastomosis at the junction of the first and second part of the duodenum after doing extensive Kocher's manoeuvre, which prevents any tension over the anastomosis.[25] The anastomosis should be well distal to the pylorus of the stomach and should not interfere significantly with pyloric function or gastric emptying.[25]

A stricture of the biliary-enteric anastomotic site is the most important complication which actually determines the long-term outcome and can occur in all types of bilioenteric anastomosis.[24] Anastomotic stricture was seen in patients with HJ who developed repeated cholangitis and needed revision surgery; none in the HD group developed a stricture. According to our experience, any anastomosis, be it in HD or HJ, should be done at the hilum or very near to the hilum for wider stoma to prevent anastomosis stricture, as described by Todani et al.[5]

We found that patients in the HD group had significantly shorter hospital stay compared to the HJ group. Another advantage of HD over HJ is that if the patient develops stricture or cholangitis in the long term, it is easier to do endoscopic intervention (endoscopic drainage or stenting) in the HD group than in the other.

During the early phase of this study, JGIHD was performed in six patients with a single case of postoperative

anastomosis leak (16.7%) without any significant short-term benefits. JGIHD is a more time-consuming and a more technically challenging procedure than either HJ or HD. As the jejunum is taken separately as graft, there are more chances of anastomosis leaks and gut viability-related complications.[29]

Conclusion

For the management of choledochal cyst, thorough clinical and other supportive laboratory and radiological investigations are needed. Symptomatic management should be done in cases of primary perforative biliary peritonitis by laparotomy followed by adequate peritoneal toileting and drainage in such cases. For patients with preoperative cholangitis, nasobiliary drainage should be the first intervention of choice. In either of these scenarios, only after proper stabilisation, should the patient be taken up for definitive operation.

For definitive management, the main aim should be complete excision of the cyst or complete excision the cyst mucosa (Lilly's technique).

For reconstruction purposes, based on the experience from this study, HD after excision of a choledochal cyst is a better alternative to HJ. It is technically easier, less complicated and less time consuming than HJ. HD requires less anastomosis, so theoretically, there are fewer chances of anastomosis-related complications. In HD, hospital stay is significantly shorter than in HJ. Chances of complications due to anastomotic stricture are also fewer in the HD group. Chance of reflux/gastritis is higher in the HD group, which was however not statistically significantly different.

None of our patients developed cholangiocarcinoma over the 30-year span of the study. However, lifetime risk for development of cholangiocarcinoma could not be evaluated, since no patients could be followed up for a lifetime.

This study in its 30-year span found no significant advantage of HJ over HD. HD should therefore be preferred, since it is easier and less time consuming to perform with theoretically fewer chances of complications.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethical approval was obtained from the Institutional Ethics Committee of Institute of Child Health, Kolkata, India (Ref: IEC/256/2021).

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Vascular rings – a case report on two varied presentations

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Summary
Vascular rings are rare congenital abnormalities of the aortic arch and branches. The ring causes varying degrees of extrinsic tracheo-oesophageal compression resulting in aerodigestive manifestations. We present two patients with atypical presentations of vascular rings. The first is a neonate with a right aortic arch, aberrant left subclavian artery and patent ductus arteriosus, diagnosed during a right posterolateral thoracotomy for oesophageal atresia and tracheo-oesophageal fistula repair. The second is a toddler presented with an impacted oesophageal foreign body. A double aortic arch causing partial oesophageal obstruction was discovered. Both patients were successfully managed by surgical division of the vascular rings.
Keywords: vascular rings, congenital abnormalities, aortic arch

Case 1

A term baby girl was delivered via normal vaginal delivery after an unremarkable pregnancy. Her birth weight was 2 510 g. Feeding intolerance was noted on day 2 of life and a coiled nasogastric tube and bowel gas in the abdomen was appreciable on a plain chest and abdominal radiograph. A diagnosis of oesophageal atresia with a distal trachea-oesophageal fistula (OA/TOF) was made. An urgent echocardiogram (ECHO) was performed as part of the preoperative assessment which reported an insignificant aortic coarctation and patent ductus arteriosus (PDA). The patient was taken for an open OA/TOF repair. The approach was via a right posterolateral thoracotomy, and on exposure of the posterior mediastinum and division of the azygos vein, an anomaly of the great vessels forming a vascular ring was visualised. Suspecting extrinsic oesophageal compression, a bronchoscopy and an oesophagoscopy were performed which confirmed the diagnosis of oesophageal atresia and distal trachea-oesophageal fistula. In order to divide the oesophageal fistula and repair the atresia, the vascular ring needed to be divided to gain exposure, however, as the anatomy was unclear at this point, division of the ring was considered unsafe and OA/TOF repair was postponed. A computed tomography angiogram (CTA) was performed postoperatively with a 3D reconstruction as demonstrated in Figure 1. This demonstrated a right-sided aortic arch (RAA) with the left carotid artery arising from it, a left-sided PDA, the left subclavian artery arising from the anterior component of the vascular ring, and an aberrant left subclavian artery (ALSA).

After medical management of nosocomial sepsis, a left thoracotomy was performed for division of the vascular ring and repair of OA/TOF by both cardiothoracic and paediatric surgery teams. The PDA was ligated proximal to the origin of the ALSA. Once the ring was divided, the TOF was identified, divided and closed, and a primary oesophago-oesophageal anastomosis was completed.

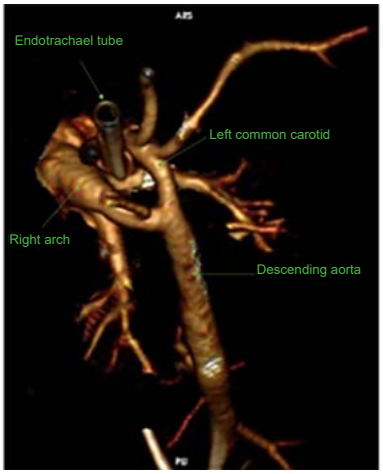


Figure 1: 3D reconstruction; right superior lateral view; a right-sided aortic arch with left side PDA and ALSA, arising from a large PDA

The postoperative recovery was unremarkable, with oral feeding initiated on day 7. Repeat cardiac evaluation excluded any intra-cardiac anomaly. The child was discharged 2 weeks after surgery, and at a 4-month outpatient follow-up was noted to be asymptomatic and growing well.

Case 2

A previously healthy 1-year 4-month old boy presented with a 10-day history of a cough and dyspnoea. On examination, the boy was irritable, tachycardic, and in respiratory distress but did not require ventilatory support. Plain anteroposterior and lateral chest radiographs demonstrated a circular radio-opaque foreign body below the thoracic inlet, positioned

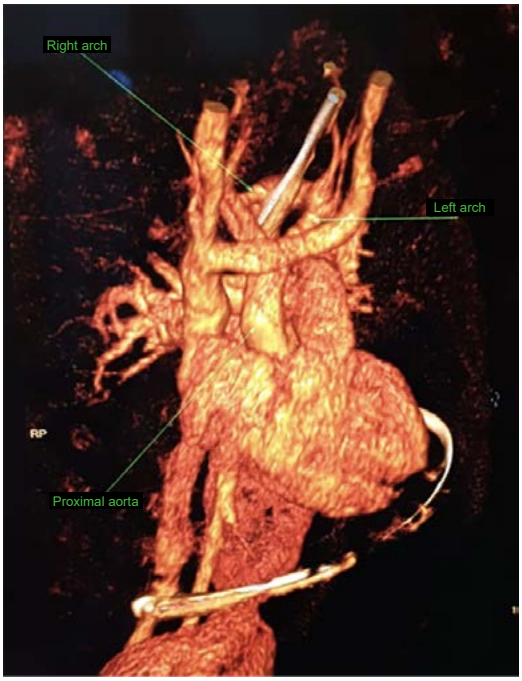


Figure 2: 3D reconstruction demonstrating right and left aortic arches

within the oesophagus. After appropriate resuscitation and initiation of systemic antibiotics, the patient underwent rigid oesophagoscopy and foreign body removal. Oesophagoscopy revealed a South African 20-cent coin (measuring 19 mm diameter), which was removed under vision. Intraoperatively, the child was difficult to ventilate and warranted further investigation. A contrasted computed tomography of the chest (CT chest) was performed. This demonstrated a double aortic arch encompassing the oesophagus and the trachea as seen in Figure 2. An echocardiogram demonstrated normal pulmonary pressures and no other structural cardiac abnormalities. A patent ductus arteriosus was not visualised.

The child was transferred to the cardiothoracic unit for definitive surgical management, this through a limited left posterolateral thoracotomy. Standard monitoring included a right radial arterial line, near-infrared spectroscopy (NIRS) as well as non-invasive blood pressure and oximetry monitoring of the left upper and lower limbs. The left arch, ductal remnant, left subclavian artery, distal right arch, left recurrent laryngeal and vagus nerves were visualised and mobilised. A test occlusion of the left arch just proximal to its confluence with the right arch was performed with no discernible change in upper versus lower limb blood pressures or NIRS readings. The left arch was then divided distal to the left subclavian artery and proximally to the area of confluence, and each end was oversewn. The ductal remnant was also divided. There was immediate compressive relief of the oesophagus and trachea. The child's postoperative course was unremarkable and at outpatient follow-up he was asymptomatic.

Discussion

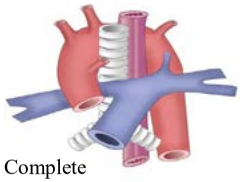
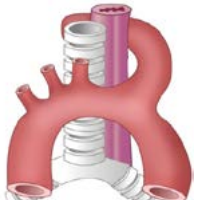
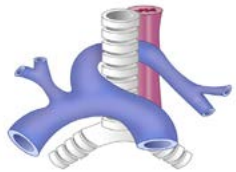
Vascular rings include a spectrum of rare anatomical malformations of the aorta resulting in either a complete or incomplete ring around the trachea and oesophagus.[1] The ring may be comprised of patent vessels, atretic vascular remnants, or ligamentous attachments.[2] The reported incidence is low and is only seen in 1–3 % of all cardiac malformations.[1]

The International Congenital Heart Surgery Nomenclature and Database Committee classified vascular rings.[3] Table I summarises the common types of complete and incomplete rings with illustration and details of the embryology, presentation, investigations and management of each.

The two cases presented above demonstrate alternative presentations of vascular rings. The incidence of an RAA in a patient with OA/TOF is about 5%.[4] Other variations in aortic anatomy, including left aortic arch with aberrant right subclavian artery, and double aortic arch, are less commonly associated with OA/TOF.[5] Neonates with long gap OA and other cardiac lesions (atrial and ventricular septal defects and tetralogy of Fallot) have an increased incidence of a coexisting vascular ring.[5] The aortic arch anatomy is assessed as part of preoperative work-up using ECHO.[6] Although ECHO is suggestive, there are limitations particularly in poor acoustic views. Laterality of the arch should be demonstrated, but the presence of an ALSA or ligamentous structures will not be clearly delineated.[2,6] The presence of the right-sided arch impacts surgical planning. Although an RAA is not a contraindication to right-sided thoracotomy approach to repair the OA/TOF, the presence of an ALSA and left-sided PDA/ligamentum arteriosum creates a complete ring.[5] Therefore, as in Case 1, these rings cannot be safely divided via right thoracotomy. Cross-sectional imaging is necessary to adequately assess patent vessels, atretic vascular elements and ligamentous attachment surrounding the trachea and oesophagus.[2] Intraoperatively, the role of bronchoscopy prior to OA/TOF repair is debated where the risk of potential decompensation must be considered against the benefit of diagnosing a proximal and distal fistula, locating the fistula and diagnosing extrinsic compression.[7] Bronchoscopy is not part of the surgical protocol for OA/TOF repair in our centre. While bronchoscopy can suggest the presence of a vascular ring, it does not inform the anatomy for surgical planning or the associated cardiac anomalies. It can, however, be useful in assessing the efficacy of ring division intraoperatively.[1] Cardiac anomalies can be a prognostic factor in patients with OA/TOF but the presence of an RAA independently is not.[4]

Older children with vascular rings present with a spectrum of trachea-oesophageal compressive symptoms.[1] Repeated visits and progression of symptoms despite adequate treatment should prompt further investigation.[8] Oesophageal foreign body impaction as an initial presentation of a vascular ring without a background history of chronic dysphagia or recurrent respiratory symptoms, as in Case 2, is rare.[9] A double aortic arch will typically present earlier in infancy due to tighter extrinsic compression.[1] However, the child in Case 2 had no history of aerodigestive complaints. The vast majority of patients will have plain chest radiographs as their first investigation. The location of the aortic arch in relation to the trachea may be helpful in suspecting a vascular ring.[10] A foreign body in the oesophagus on chest radiograph can be a secondary presentation of a vascular ring.[2] It is important to carefully assess the foreign body and risk of perforation, particularly in the presence of a vascular ring, because of the risk of catastrophic bleed from aorto-oesophageal fistula once removed.

Table I: Vascular rings

Anomaly	Embryology	Age at presentation	Symptoms	Chest radiograph	Contrast swallow	Bronchoscopy	Management	Other information
Double aortic arch  Complete	Persistence of fourth branchial arches bilaterally Right aorta fails to regress	1–3 months	Respiratory distress Stridor Brassy cough Swallowing difficulties Apnoeic episodes Cyanotic spells Recurrent respiratory tract infections Failure to thrive	AP (anterior posterior) view Widened mediastinum Lateral view Compression of trachea	AP view Bilateral indentation Lateral view Posterior indentation	Bilateral tracheal compression-both sides pulsatile	**Surgery** Division of smaller arch (usually left) and PDA ligation. Left PDA: Left thoracotomy Right PDA: Right thoracotomy	Commonest type Uncommonly associated with other cardiac anomalies
Right aortic arch with left sided PDA/ ligamentum arteriosum  Complete	Left fourth arch regresses (between the aorta and left subclavian) Remnant of left fourth arch-Kommerell's diverticulum	6–12 months	Respiratory distress Swallowing difficulties (detected when introducing solids)	AP view Tracheal deviation to the left	AP view Bilateral indentation Right > left Lateral view Posterior indentation	Bilateral tracheal compression-only right side pulsatile	**Surgery** Division of ductus or ligamentum arteriosum via left thoracotomy	5–10% association with cardiac anomalies 5% of OA/TOF have coexisting RAA Aberrant left subclavian artery may arise from Kommerell's diverticulum
Anomalous innominate artery  Incomplete	Aberrant innominate artery can arise from diverticulum	< 2 years	Respiratory distress Cyanosis Stridor Apnoeic episodes Hyperextension of head in an attempt to splint trachea	Lateral view Tracheal compression anteriorly	Normal	Anterior compression-pulsatile	**Non-surgical** Often become asymptomatic > 2 years **Surgery** Innominate artery or aortopexy procedure pexying to sternum	Only for surgical intervention if tracheal lumen stenosis is > 70%
Aberrant right subclavian artery  Incomplete	Regression of the right fourth arch and proximal right dorsal aorta Aberrant right subclavian forms from the persistence of the seventh intersegmental artery	Seventh and eighth decades of life	Calcification of the aberrant vessel leads to dysphagia lusoria	Normal	AP view Oblique right-sided impression	Normal	**Surgery** Hybrid approach: thoracic endovascular aortic repair in conjunction with carotid subclavian artery bypass	Known as arteria lusoria Lusoria means 'freak of nature" in Latin Dilated root of the aberrant right subclavian is Kommerell's diverticulum
Pulmonary artery sling  Incomplete	Left pulmonary artery (LPA) originates from the right pulmonary artery (RPA) and encircles at the level of the carina	< 1 year	Respiratory distress Stridor Apnoeas Cyanotic spells Recurrent respiratory tract infections	AP view Right-sided air trapping or atelectasis Lateral view Anterior bowing of trachea and right main bronchus	Lateral view Anterior indentation between carina and oesophagus	Displacement of tracheal to left and compression of right main bronchus	**Surgery** Left pulmonary artery is reimplanted from its right-sided origin to the main pulmonary artery	30–40% association with tracheal rings

Conclusion

Vascular rings are rare congenital anomalies. These cases represent the spectrum of presentation of vascular rings and the importance of a high index of suspicion. They can coexist with commonly managed paediatric surgical conditions where an atypical clinical presentation may be demonstrated, and the presence of the vascular ring may complicate the clinical course and surgical intervention. A vascular ring in a neonate, without an associated OA/TOF is likely to be a tight complete ring like a double aortic arch and may only present with unexplained respiratory distress. In the case of a coexisting OA/TOF as in Case 1, the purpose of a preoperative ECHO is to exclude an RAA which can lack sensitivity. Once the arch was identified intraoperatively, the appropriate and safe decision for further cross-sectional imaging before further action was made. In Case 2, although the child was unusual as he was previously well, the small size of the impacted foreign body and persistent ventilatory difficulties after removal should have alerted us to the possibility of extrinsic compression earlier. Patients with vascular rings who have an early diagnosis, appropriate investigations and timeous surgical intervention are likely to make a full recovery.

Conflict of interest

The authors declare no conflict of interest.

Ethical approval

Ethical approval was obtained from the University of the Witwatersrand Human Research Ethics Committee (Medical): M1811139.

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PAEDIATRIC SURGERY

South Africans abroad – international fellowship experience amongst South African paediatric surgeons

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Background: South African paediatric surgeons complete broad-based general training at various centres throughout South Africa. Electives at units elsewhere in the country or abroad are not traditional, and as yet there are no formal sub-specialty training programmes. International fellowships offer South Africans the opportunity to experience foreign healthcare systems, spend time in high-volume sub-specialty units, and add diversity to their own skill set and to the unit to which they return.
Methods: Eleven paediatric surgeons known to have completed international fellowships were sent a questionnaire regarding their fellowship experience. They were specifically asked about their fellowship's clinical and academic benefits, the impact of the fellowship on their career progression, and financial implications of the fellowship.
Results: All respondents felt that their fellowship had been clinically advantageous. Most found that it had been academically beneficial. Formal career progression as a result of the fellowship was difficult to quantify and, overall, there seemed to be no financial advantage to completing an international fellowship.
Conclusion: International fellowships can be financially challenging and may offer no concrete career progression. However, they are clinically and academically advantageous and are perceived to be generally beneficial to those who undertake them.
Keywords: paediatric surgery, fellowship, global surgery

Background

Described as 'true generalists' by Michael Dinner in the 1970s,[1] South African paediatric surgeons undergo broad-based clinical training at various academic centres throughout the country. By the time they sit their exit college exams, trainees should have had exposure to the majority of paediatric surgical conditions and are expected to function largely as independent surgeons once registrar training is complete. Currently there are no formalised paediatric surgical sub-specialty fellowships in South Africa.

Paediatric surgeons may elect to participate in overseas fellowships during or after their registrar training. Although these fellowships are not formally recognised as additional training by the Health Professions Council of South Africa (HPCSA), they may confer some benefits to those who choose to undertake them, as well as to the surgical units to which these clinicians return.

In this short report, the authors detail the results of a survey sent out to South African paediatric surgeons who completed fellowships abroad. The purpose of the survey was to gain clarity on their experiences and their recommendations for those considering overseas fellowship training in future.

Methods

A REDCap survey was sent out to acquaintances of the authors known to have completed overseas fellowships in various areas of paediatric surgery. This was a sample of convenience and does not represent all South African paediatric surgeons to have ever undertaken international fellowships. The survey (Appendix A) was administered for the purposes of a lecture given at the 2022 University of the Witwatersrand (Wits) Global Paediatric Surgical Conference by the first author and aimed to gather information on several aspects of the fellowships, including their perceived usefulness from a clinical and academic perspective, their perceived role in career progression and their financial implications. Respondents were asked to comment on whether or not they would recommend an international fellowship to trainees. Some demographic data were also obtained.

Results

There were 11 respondents, aged between 39 and 81 years. Seven of the respondents were male. One respondent is fully retired, another is retired from state practice but still assists in the private sector: both of these respondents previously served as heads of department (HOD). The remaining nine respondents are all currently practicing as paediatric surgeons, three of whom are currently in HOD positions. Three respondents currently reside outside of South Africa. Respondents had completed fellowships in colorectal surgery, hepatobiliary surgery, oncology, thoracic surgery, paediatric transplant, paediatric urology, paediatric burns, and general paediatric surgery. Countries trained in included the United Kingdom (n = 5 respondents), Canada (n = 3),

Australia (n = 3), the United States (n = 2), and Germany (n = 1). Two respondents had participated in more than one fellowship: these fellowships were undertaken in separate countries and in different sub-specialties. Fellowships ranged in duration from 6 to 24 months.

Clinical benefits

All respondents reported finding their fellowships clinically beneficial. Factors such as the opportunity to work in high volume units with exposure to rare pathologies under the supervision of internationally renowned surgeons were listed as being of particular benefit. Respondents also reported that being exposed to new surgical techniques, alternative ways of thinking and foreign healthcare systems were valuable. Also valuable was the experience of working within surgical teams that contained members with diverse skill sets, including staff members not always found in South African units, such as nurse practitioners, specialist nurses and surgeon scientists, and observing the way these diverse teams contributed to holistic peri-operative care and follow-up.

Academic benefits

Nine out of eleven respondents reported that their fellowships were academically advantageous, with two reporting that they were not. Respondents reported being involved in work that led to multiple publications, being exposed to postgraduate teaching programmes that allowed them to sit international board exams and undertaking research that ultimately led to higher degrees such as a Master of Medicine (MMed degree). Several reported that they are still involved in collaborative work with the unit where they undertook their fellowship. Respondents who completed shorter fellowships (< 1 year) reported not having enough time to complete research projects whilst abroad, but nevertheless found the experience to be useful in their future academic endeavours. One respondent stated that the focus of their fellowship had been clinical rather than academic.

Career progression

All 11 respondents felt that their fellowship had contributed to career progression. This was difficult to quantify formally, but they stated that the fellowship gave them clinical confidence and 'improved [their] standing' amongst colleagues and 'kick-started' elements of their academic career. Four out of five respondents with current or previous HOD experience stated that their own fellowship experience contributed to their appointment as HOD, and all five stated that fellowship experience would count favourably when considering candidates for consultant positions within their units.

Financial implications

Nine out of eleven respondents reported that their fellowship was not financially advantageous. Several respondents reported that the fellowship resulted in a financial loss, whilst others reported that they ultimately 'broke even'. Two respondents recommended that surgeons considering fellowships attempt saving prior to the experience. No respondents commented on the implications of fellowship training for future earnings.

Recommendations and other considerations

Three respondents no longer live in South Africa: only one of these reported that the fellowship contributed directly to their decision to emigrate. One respondent (currently an HOD at an academic centre in South Africa) stated that they believe that posts should be created in South Africa in order to encourage fellowship-trained surgeons to return to the country.

All five current or past HODs stated that they would recommend pursuing an international fellowship to their own trainees, as did the majority of other respondents. Two respondents felt ambivalent about recommending overseas fellowship to colleagues.

Conclusion

Paediatric surgery is a relatively young discipline in South Africa; it was first recognised by the HPCSA as a formal sub-specialty of general surgery in 1984, and then converted to an independent specialty in 2007.[1] The HPCSA register currently lists 47 active paediatric surgeons practicing in South Africa, where there are around 20 million children under the age of 18. This scarcity of paediatric surgeons is well documented[2] and contributes to high case volumes at tertiary centres. Empiric evidence showing that high case volumes contribute to improved outcomes is conflicting,[3,4] but nevertheless local and international trainees and qualified surgeons have benefitted from working at South African centres and learn from the diverse pathology represented at these various academic institutions.[5-7] This trend continues throughout sub-Saharan Africa.[8]

Despite the diversity and high volume of pathology, paediatric surgical training in South Africa can be somewhat limited in terms of exposure to diverse ideas, protocols and methods due to the fact that many academic centres may only have one or two affiliated paediatric surgical units, with scanty senior staffing due to overall low numbers of specialists. South African paediatric surgery trainees do not traditionally undertake electives at other units around or outside of the country. Registrars are thus often trained in environments that might be relatively isolated without much external senior thoroughfare.

Fellowship experience is potentially valuable for the 'true generalists' and aspiring sub-specialists alike. Based on the experience of surgeons who have undertaken fellowships, electives and fellowships both abroad and within South Africa are perceived to offer the opportunity for trainees and surgeons to expand their surgical exposure, to learn about holistic peri-operative care, and to bring new ideas and general skills back to their home base. They may also present the opportunity to have learning reinforced, when fellows discover that their base-training leaves them more than adequately equipped to work and thrive in a foreign unit. Sub-specialty training has not yet been formalised within the South African paediatric surgery context, but there is a trend in some centres toward informal sub-specialisation as some consultants develop special interests. A year or two in an international, high-volume sub-specialty unit may be useful for individuals wishing to focus their practice in a certain area. As South African units themselves develop their own high-volume sub-specialist units, the possibility of local sub-specialty fellowships should be considered and perhaps eventually formalised. As things currently stand, there may be significant benefit to encouraging registrars to

participate in exchange programmes with other units within South Africa.

Furthermore, cross-border relationships created by international fellowships and electives create the opportunity for long-lasting and repeated collaborations between centres in the global North and global South. These collaborations are valuable from an academic point of view, allowing for joint participation on multi-centre research projects and allowing for sharing of resources. They are also clinically valuable, paving the way not only for ongoing clinical discussion but also establishing future pathways for fellows who wish to travel to units outside of their own. Just as there are perceived benefits for trainees from the global South completing fellowships in the global North, so too are there benefits to trainees from the Northern hemisphere spending time in the global South. The benefits to the trainees themselves are well documented[7] and anecdotally these fellows add value to the units they visit when they bring in new ideas from their home units.

It must be acknowledged that fellowships are financially challenging and need to be fitted in around other non-work-related life events. This, coupled with the fact that it is difficult to formally quantify their contribution to career progression within South Africa, may make them unattractive to many. That said, surgeons who have undertaken fellowships themselves believe them to be clinically and academically advantageous and past and present HODs surveyed for this paper all recommend international fellowships and view fellowship experience as desirable in potential employees. It also bears mentioning that overseas fellowships may contribute to the attrition of South African paediatric surgeons, who are an already scarce resource, and the creation of attractive posts for returning fellows may help to mitigate this.

Limitations of this report include the fact that respondents were limited to individuals known personally to the authors, and that the findings all represent personal opinions. A broader investigation into the implications of international fellowships for both trainees and the discipline of paediatric surgery in South Africa as a whole may be useful.

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Conflict of interest

The authors declare no conflict of interest.

Ethical approval

Ethical approval for this article was not sought. Interviewees responded to a survey at the first author's request and gave written permission for their responses to be collated into this short report.

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Appendix A

International Paediatric Surgery Fellowships

The questionnaire below intends to collate some of the experiences of South African Paediatric Surgeons who undertook international fellowships. The results of this survey will be used in a discussion session at the 2022 Wits Paediatric Surgical Conference.

Thank you for your time and participation.

Please enter your initials	_____
In which year were you born?	_____
Have you ever been a Head of Unit/Departmental Chief?	◯ Yes ◯ No
Do you feel your fellowship training contributed to your appointment to this position?	◯ Yes ◯ No ◯ I'm unsure or undecided
Would fellowship experience count favourably when you were making a decision about appointing a new consultant in your department?	◯ Yes ◯ No ◯ I'd like to give a longer answer
Tell me more about the value of fellowship experience when interviewing candidates for new consultant posts	_____
Would you recommend pursuing an international fellowship to trainees graduating from your division?	◯ Yes ◯ No
Do you still live in South Africa?	◯ Yes ◯ No
Did your fellowship contribute to your decision to move out of South Africa?	◯ Yes ◯ No ◯ I'd like to give a longer answer
Tell me more about your decision to move out of South Africa	_____
In which country/countries did you do your fellowship?	_____
In which field(s) did you do your fellowship?	_____
How long was your fellowship?	_____
Was your fellowship clinically advantageous?	◯ Yes ◯ No ◯ Neither advantageous nor disadvantageous ◯ I'm still undecided

If you like, please tell me more about the clinical aspects of your fellowship	_____
Was your fellowship academically advantageous?	◯ Yes ◯ No ◯ Neither advantageous nor disadvantageous ◯ I'm still undecided
If you like, please tell me more about the academic aspects of your fellowship	_____
Was your fellowship financially advantageous?	◯ Yes ◯ No ◯ Neither advantageous nor disadvantageous ◯ I'm still undecided
If you like, please tell me more about the financial aspects of your fellowship	_____
Did your fellowship contribute to your career progression?	◯ Yes ◯ No ◯ I'd like to give a longer answer
If you like, please tell me more about how your fellowship impacted about your career progression.	_____
Would you recommend pursuing an international fellowship to other paediatric surgeons?	◯ Yes ◯ No ◯ My answer would not be a simple 'Yes' or 'No'
Do you have any other thoughts or comments about undertaking an international fellowship in Paediatric Surgery?	_____

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PAEDIATRIC SURGERY

Podcasting with Discover Paediatric Surgery

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Background: Digital advancements combined with an exponential increase in published medical literature has led to the development of novel forms of medical education. Medical podcasts are one example of innovative education tools on the rise worldwide. Discover Paediatric Surgery is a medical podcast produced in Johannesburg that tackles some common and fundamental topics encountered within the paediatric surgical field.
Methods: Discover Paediatric Surgery was conceived of, recorded and published by the first author (AG). For the purposes of this audit, anonymous listener data for the podcast was reviewed retrospectively to determine geographic reach and the most popular subjects.
Results: Nineteen episodes were published between March 2018 and February 2019. At the time of writing, there have been 14 612 downloads by 6 182 unique listeners, with ongoing downloads occurring over time. Median downloads per episode is 668 (range 427–2 061). Podcast listeners originated from 108 countries, the majority of whom were based in South Africa, the US and the United Kingdom.
Conclusion: There is a demand for relevant and accessible medical podcasts dealing with common topics in paediatric surgery. Podcasting is a novel way to deliver medical education, which allows on-the-go learning which can be equitably accessed. The impact of medical podcasting on patient care and outcomes is yet to be determined.
Keywords: podcast, e-learning, medical education, continued medical education, paediatric surgery, registrar training, global surgery

Introduction

The exponential increase in the availability of medical information over the past decade together with major advances in the field of accessible digital technology has changed both the landscape and the expectations for medical trainees and qualified healthcare practitioners. Gone are the days of professors sitting in the library, reading the latest medical journals in hard copy, whilst students rummage through catalogues to find heavy textbooks to study from for upcoming evaluations. Now, there is near-universal access to electronic educational resources. Many of these are traditional resources such as textbooks and journals that have been converted to a digital format, but as Web 2.0 has become a part of daily life, so have its applications and platforms become integral to the transfer of medical information. The authors confirmed this transition among the delegates at a Paediatric Surgery Global congress in 2022, held in South Africa, where attendees were polled regarding their methods for acquiring medical information. Ninety per cent of attendees routinely utilised electronic textbooks, 87% used web searches for on-the-go medical information updates on a daily basis, and all participants confirmed that they listened to medical podcasts.

Podcasts are recorded audio content that is distributed via a streaming service and is usually accessed through a web-linked device such as a smartphone or home internet speaker. Medical podcasting is a well-established endeavour with content that is intended for undergraduate,[1] postgraduate and continued medical education.[2,3] There are podcasts available for a broad range of medical specialties,[2,4-7] including paediatric surgery. The utility of podcasts as educational tools in medical training has been extensively studied and evaluated,[3,5-7] and many major medical journals and medical associations produce their own podcasts or run editorials recommending podcasts to listen to.[8,9] Podcasts are also being introduced into formal medical curricula[10] and their usage increased significantly during the COVID-19 pandemic.[11]

This paper is a retrospective review of the listenership characteristics of a South African-developed paediatric surgery podcast called Discover Paediatric Surgery. The first author (AG), who works as a consultant paediatric surgeon at a tertiary academic centre, noted that local paediatric surgery trainees, faced with a constant onslaught of new information via journals and textbooks, seemed to be having significant challenges assimilating knowledge around topics core to their specialty. It is also well understood that the majority[12] of paediatric surgical procedures performed in Africa are undertaken by general surgeons with minimal formal training within this sub-speciality. With this in mind, the podcast Discover Paediatric Surgery was developed to provide an accessible and up-to-date educational tool for people practicing paediatric surgery. Published in a format that is available on widely used platforms, Discover Paediatric Surgery can be accessed by anyone with a mobile or desktop device and an internet connection.

Methods

Discover Paediatric Surgery was self-produced by AG. AG is responsible for recording and editing each episode, after

which it is uploaded to Captivate (Captivate Audio Ltd, United Kingdom, 2022) which then distributes the podcast to platforms such as Apple Podcasts (Apple Inc, United States, 2022) and Stitcher (Stitcher, 2022). Listeners are able to download the podcast through these platforms at no cost other than the data costs determined by their service provider. Captivate then provides anonymous listener data and metrics to the podcast producer, including listener location, the device and operating system used to access the podcast, and the number of listens over time. The podcast covers various topics in Paediatric Surgery. Each episode features a guest: these guests are professional acquaintances of AG and well-regarded within their subspecialist fields. AG takes responsibility for the final quality of the podcast and the data contained within. Discover Paediatric Surgery is recorded in English which has implications for its listenership.

Results

Discovery Paediatric Surgery has published 19 episodes since April 2018 with each episode being between 20 and 54 minutes in length (excluding the introduction podcast). Some topics are broken down into two parts.

Podcast topics fall into eight broad categories: hepatobiliary conditions (4 episodes); urology (3 episodes); thoracic conditions including oesophageal atresia and trachea-oesophageal fistula (3 episodes); neonatal and infantile conditions (3 episodes); upper gastro-intestinal conditions (1 episode); colorectal conditions (1 episode); neurosurgical trauma (1 episode); and miscellaneous topics including sedation and analgesia (2 episodes). Guests included 8 South African clinicians (7 paediatric surgeons, 1 nephrologist, 1 anaesthesiologist) and five internationally-based surgeons and urologists, all of whom currently work in Europe. Of the South African clinicians, one was a paediatric surgery trainee at the time of recording.

Overall, Discovery Paediatric Surgery has had 14 612 downloads, and 6 182 unique listeners in 108 countries with an episode median download number of 669 per episode. The most downloaded episode is Discover Hirschsprung's Disease (2 061 downloads), which also makes colorectal surgery the most popular category. Forty-two per cent of downloads come from South Africa, 15% from the United States of America, 12% from the United Kingdom and 7% from Australia (Figure 1). Users downloaded the podcast

Figure 1: Choropleth map of downloads origins

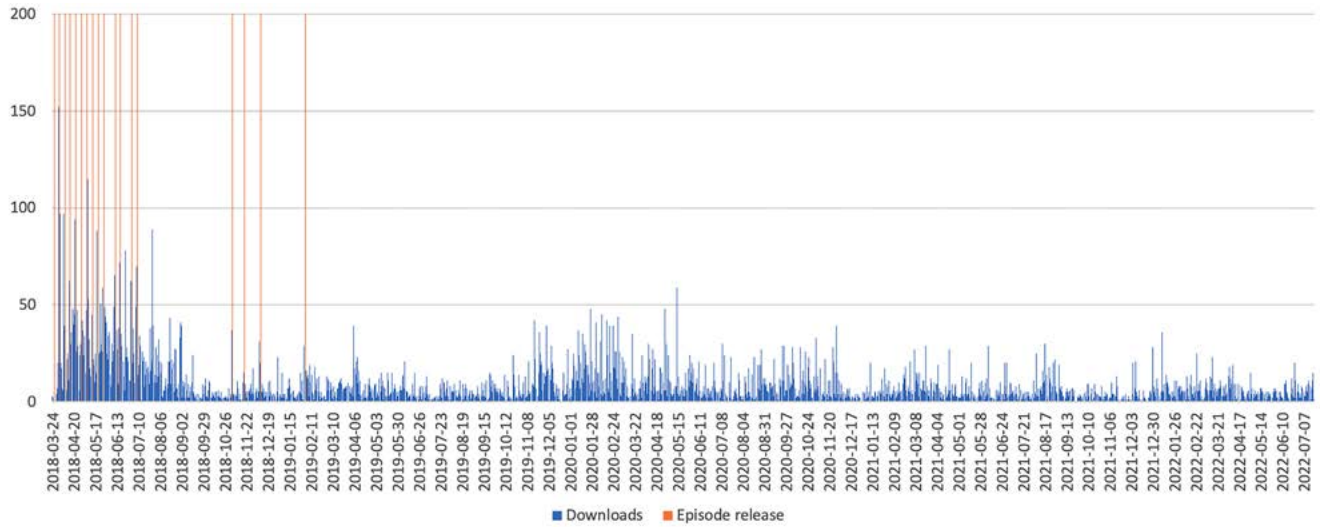

Figure 2: Daily download numbers

using a mobile device 47% of the time, using either a mobile app or browser. Episodes were downloaded most frequently immediately after release, but longitudinal data show ongoing downloads over time, likely indicating a sustained interest in the topics addressed (Figure 2).

Discover Paediatric Surgery has an average rating of 4.8 stars on Apple Podcasts.

Discussion

Access to safe surgery is a major challenge in low- and middle-income countries (LMICs), where over 5 billion people do not have reliable surgical access. This results in an estimated 17 million avoidable deaths per annum.[13] The lack of specialist paediatric surgeons in Africa necessitates that the majority of paediatric surgical procedures be done by general surgeons with little to no specific training in paediatric surgery.[12] There is thus a need to increase paediatric surgical knowledge and increase awareness of paediatric surgical conditions amongst general surgeons compelled to treat children with surgical disease. Additionally, paediatric surgical trainees and specialists are challenged by overwhelming volumes of literature which cannot all be parsed in the relatively scarce non-clinical time available each week. Knowledge-transfer platforms which are succinct, convenient to access and relevant are now essential to medical education, and it was with this in mind that Discover Paediatric Surgery was developed.

The term 'podcasting' was coined in 2004 when a new form of 'online radio' which combined the 'intimacy of voice, the interactivity of a weblog, and the convenience and portability of an MP3 download' was described.[14] Made possible by the increasing availability of cheap or free audio production software and the boom in portable, personal devices such MP3 players, iPods, smartphones and home internet speakers, podcasting took off in the mid-2000s and is an industry that grows annually. It is estimated that in 2022 podcast listeners worldwide number in the hundreds of millions.[15] In major South African metros in 2021, 48% of the population aged 15 and up was aware of podcasts, and 36% of this population listened to podcasts.[16]

The use of audio recordings to facilitate medical teaching has been described since the early 1960s, when magnetic tape recordings were first made of lectures and talks. These recordings where sometimes combined with transparencies that could be projected on cue whilst listening to the lectures.[17] Although lectures, tutorials and reference books remain central to the study of medicine, the impact of various media including websites, weblogs, interactive multimedia journal articles and videos hosted on platforms such as YouTube is significant. The use of medical podcasts is also increasing over time and they are used extensively by undergraduates and postgraduates who listen to them whilst engaging in other activities such as driving, completing chores and exercising.[1,5] A recent scoping review found that podcasts are valued for their efficiency and combined entertainment and educational ('edutainment') value, that they are not inferior to traditional teaching methods in terms of knowledge retention, and that they possibly effect behaviour and practice change in medical students, registrars and specialists.[10] To date, however, there is little evidence to show that podcast use changes patient outcomes.[6,10]

When discussing podcasts with users, certain features are deemed desirable. These include a podcast length of somewhere between 15 and 30 minutes, depending on the topic,[1,3,6,10] the incorporation of dialogue that creates a conversational tone, the use of credible source material, summarised key points and the explanation of acronyms.[10] Quality indicators for blogs and podcasts used in medical education include credibility and transparency; clear distinctions between facts, opinions, content and advertisements; citation of references and factual consistency with these; as well as good design and functionality that allows the resource to be available to learners with standard equipment and software.[7]

The need for additional paediatric surgeons in LMICs is well documented, but the likelihood of producing the required number of specialists within the next decade is unrealistic. We can, however, strive to educate around commonly encountered paediatric surgical conditions to create a solid workforce of well-qualified paediatric surgeons. Podcasts such as Discover Paediatric Surgery are free, easily accessible, and perfectly positioned as a useful additional resource to achieve these goals. The penetration and relevance of a podcast such as Discovery Paediatric Surgery in LMICs may be improved by interviewing guests from some of these countries, by recording episodes in regional languages, and by advertising the podcast in local general surgical journals and on social media platforms.

Podcasting does have its limitations, chief among these being the time and financial resources required to undertake such an endeavour, which includes significant technical upskilling in an area not focused on during surgical training. To date there is also minimal data to show the impact of podcast-delivered education on patient outcomes, which is a topic for further research.

Conclusion

In summary, download and listenership data for Discover Paediatric Surgery, a podcast that focuses on common topics in paediatric surgery, show that there is enthusiasm for this type of content both in South Africa and abroad. Podcasts provide accessible and up-to-date discussion via widely utilised platforms and are likely to form a significant component of medical education in the near future.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

The authors declare that this submission is in accordance with the principles laid down by the Responsible Research Publication Position Statements as developed at the 2nd World Conference on Research Integrity in Singapore, 2010.

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PAEDIATRIC SURGERY

A topographical description of the recurrent laryngeal nerve in paediatric cadavers

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Background: The risk of injury to the recurrent laryngeal nerve (RLN) during thyroidectomy is higher in children. The aim of the study was to document the anatomy of the RLN in paediatric cadavers.
Methods: A midline incision from the mandible to the sternal notch was used to expose the structures. Exposed tissues were photographed at 15–30 cm and 45º angle to the neck. The relationship of the RLN to the inferior thyroid artery (ITA) and trachea-oesophageal groove (TEG) was documented. Extra-laryngeal branching of the RLN was noted. The angles which the RLN made relative to the longitudinal axis of the TEG were documented.
Results: A total of 31 cadavers with a mean age of 39.7 ± 51.7 post-natal weeks, totalling 61 RLNs were studied. Of the right RLNs, 66.7% were within 0–15º of the TEG compared to 96% on the left. Around 66.7% (20/30) of the RLNs passed anterior to the ITA on the right compared to 58.1% (18/31) on the left. Extra-laryngeal branching of the RLN was observed in 29.0% of specimens and the greatest number of branches noted were two.
Conclusion: The RLN on the left side ran within 0–15º of the TEG in 96% of the cases. Around 68% of RLNs on the right side passed anterior to the ITA. Close to 30% of the RLNs gave branches. The course of the paediatric RLN is highly variable and findings on the contralateral side should not be relied on to guide dissection on the opposite side during thyroidectomy in children.
Keywords: recurrent laryngeal nerve, inferior thyroid artery, trachea-oesophageal groove, extra-laryngeal branches

Background

Thyroidectomy is rarely performed in children and thyroid cancer is among the more common indications.[1-3] The majority of children with thyroid cancer present when the cancer has already metastasised to lymph nodes, necessitating total thyroidectomy and cervical lymph node dissection.[4] Furthermore, thyroid cancer in children is likely to be multicentric.

Total thyroidectomy with or without cervical lymph node dissection is complex and is associated with a higher risk of complications such as injury to the recurrent laryngeal nerve (RLN). The complications associated with thyroidectomy occur even when the procedure is performed by experienced surgeons, and the risk of complications is higher when performed in children.[5] This is because of the miniature size of anatomical structures and their proximity in the neck.[1] The possible mechanisms of injury of RLN during thyroidectomy include traction injury, clamping, thermal damage, ligation, transection, suctioning and ischemia.[6]

Strategies which are used to reduce the likelihood of injury to the RLN during thyroidectomy include its identification followed by meticulous tracking to where the nerve enters the larynx[6] and intraoperative neuro-monitoring.[7] The landmarks which are used for the identification of the RLN during thyroidectomy are the inferior thyroid artery (ITA), trachea-oesophageal groove (TEG) and the ligament of Berry. However, the anatomy of the RLN is highly variable and asymmetrical in adults. The inconsistencies in the anatomy of the RLN include the existence and patterns of extra-laryngeal branching. There is a geographical difference in the anatomy of the RLN.[8] The key to safely conducting thyroid surgery in children remains a good knowledge of the anatomy of the RLN, which unfortunately is known to be highly variable in adults.[9]

Thyroidectomy in children should be done by a team of surgeons that includes a high-volume thyroid surgeon and a paediatric surgeon. It is rare to find surgeons who would have reached the prescribed minimum number based on the number of thyroidectomies which they perform solely in children. The requisite 30 thyroidectomies per year, which are necessary to meet the threshold for a high-volume thyroid surgeon, are not easily achieved by most adult thyroid surgeons and paediatric surgeons as thyroid pathologies are rare in children.[10] The technique of thyroidectomy in children is therefore often extrapolated from the experience gained from thyroidectomies done in adults.[10]

There is a paucity of information regarding the anatomy of the RLN including the prevalence of anatomical variations in children.[8] The aim of this research was therefore to study the anatomy of the RLN in paediatric cadavers.

Materials and methods

Paediatric cadavers were sourced from the School of Anatomical Sciences of the University of the Witwatersrand

and the Department of Anatomical Sciences at the University of Pretoria. These were then dissected and photographed.

A total of 31 cadavers with a mean age of 39.7 ± 51.7 post-natal weeks were studied. One RLN on the right could not be identified. This was ascribed to the poor state of the cadaver due to decomposition. The possibility of a non-recurrent laryngeal nerve was entertained after completion of the study. Overall, 61 RLNs were identified and studied. A midline incision was made from the mental process of the mandible to the sternal notch. The skin was reflected to expose the platysma followed by opening of the investing layer of fascia to expose the supra- and infrahyoid muscles. These muscles were then reflected to reveal the carotid sheath and its contents. The sternoclavicular joint was disarticulated to visualise the point of recurrence of the RLN. The internal jugular vein was removed to achieve better visibility.

The RLN was followed as it coursed in the TEG. The relation of the RLN to the ITA and TEG on either side of the neck was documented. Both the course and relations of the RLN, including the angle they made in relation to the TEG, were studied. The angle which the RLN made with the TEG was categorised into four groups: $X < 15°$; $15° < X < 30°$; $30° < X < 45°$ and $X > 45°$. The terminal end of the RLN was examined and the presence or absence of branches, including the number of branches, was documented.

Photographs were taken at 15–30 cm from the specimen and at a 45° angle to the neck using a high-resolution Nikon® camera and analysed with the open-source software ImageJ® (National Institutes of Health in Bethesda, Maryland, United States of America). A standard 1 mm scale bar was applied to all the pictures. Following scaling, the software programme ImageJ® calculated pixel differences between two selected points and translated them into accurate metric measurements.

The collected data was entered onto a spreadsheet in Microsoft Excel 2013 (Microsoft Corporation). STATA® version 14 statistical software was used for analysis. Percentages were used to summarise categorical data. Measurements derived from the photographs were compared using a student's t-test. The topography of the RLN including its relationship with the ITA and TEG and branching pattern were compared using a chi-square test to assess respective associations. Correlation and regression analysis were done to assess the regularity and the predictability of the RLN relationship with both the TEG and ITA, and its branching patterns.

Results

The mean diameter of the RLN in the paediatric cadaveric population was 0.74 ± 0.28 mm on the left and 0.72 ± 0.37 mm on the right. The analysis of the mean diameter of the RLN in paediatric cadavers was not categorised according to gender. All the RLNs which could be identified were recurrent, and one nerve turned inferior to a patent ductus arteriosus (Figure 1).

On the right side, 43.3% (13/30) of the RLN followed the course of the TEG, but 56.7% (17/30) deviated from the TEG creating an angle ranging from 6.7–44° in size. On the left side, 90.3% (28/31) of RLNs were intimately related to the TEG, while 9.7 % (3/31) deviated from the TEG. In total, 96.8% (30/31) of the RLNs on the left and 66.7% (20/30) on the right side were found coursing within 0–15° angle to the TEG (Table I and Figure 2).

Table I: Table showing the percentage distribution of the angular course of the recurrent laryngeal in the trachea-oesophageal groove

Angle	Left (%)	Right (%)
< 15°	96.8% (30/31)	66.7% (20/30)
15–30°	0% (0/31)	23.3% (7/30)
31–45°	3.2% (1/31)	10% (3/30)
> 45°	0% (0/31)	0% (0/30)

Symmetry of the angular course in relation to the TEG was observed in 40% (12/30 pairs) of RLNs. A student's t-test revealed that the difference in the anatomical relationship of the RLNs with the TEG between the sides was statistically significant with a *p*-value of 0.017. The RLNs passed anterior to the ITA in 66.7% (20/30) on the right side compared to 58.1% (18/31) on the left side ITA (Table II).

Table II: Percentage distribution of anatomical relation between the recurrent laryngeal nerve and the inferior thyroid artery

Parameter	Anterior to ITA	Between branches of ITA	Posterior to ITA
Right	66.7% (20/30)	13.3% (4/30)	20.0% (6/30)
Left	58.1% (18/31)	3.2% (1/31)	38.7% (12/31)
Overall	62.3% (38/61)	8.2% (5/61)	29.5% (18/61)

Figure 3 and Figure 4 show the RLN passing anterior to the ITA on the right and left side of the neck.

Extra-laryngeal branching of the RLN was observed in 29.5% (18/61) of the cases. In 5.6% (1/18) of these cases, bifurcation of the RLN was observed bilaterally. The difference in the branching of the RLN was statistically significant ($p < 0.05$; value = 0.008266). The highest number

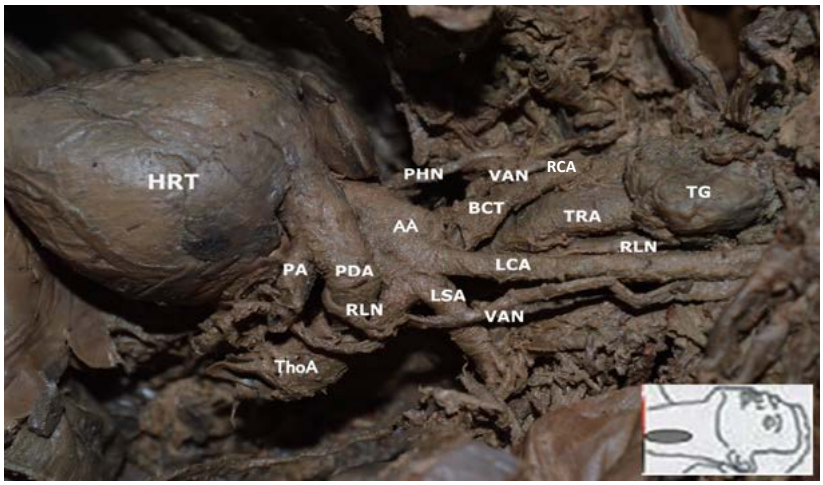

Figure 1: Photograph showing the variant pattern of the recurrent laryngeal nerve recurring below a patent ductus arteriosus on the left side

RLN – left recurrent laryngeal nerve, TG – thyroid gland, TRA – trachea, HRT – heart, PA – pulmonary artery, PDA – patent ductus arteriosus, ThoA – thoracic aorta, LCA – left common carotid artery, LSA – left subclavian artery, BCT – brachiocephalic trunk, RCA – right common carotid artery, PHN – phrenic nerve, VAN – vagus nerve

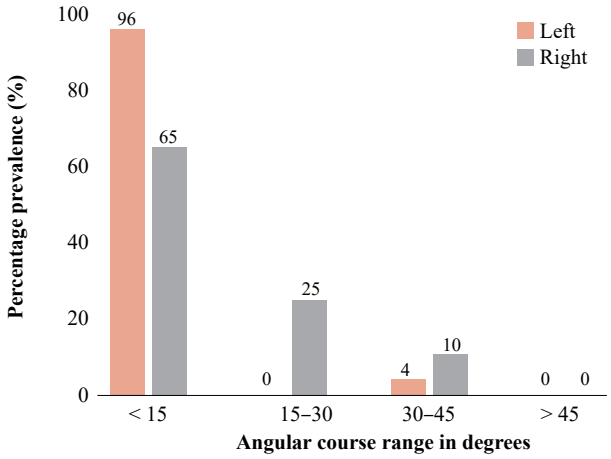

Figure 2: Percentage distribution of the angular course of the RLN in the TEG

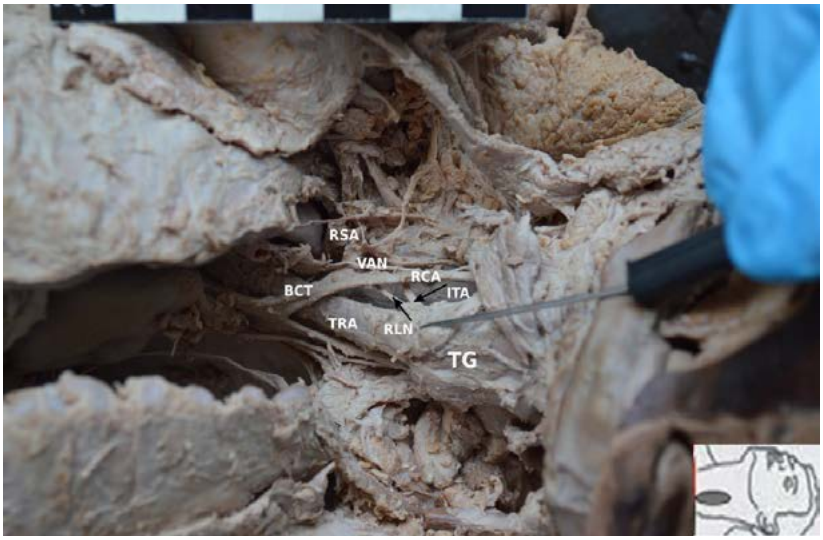

Figure 3: Photograph showing the RLN coursing anterior to the ITA on the right side

RLN – recurrent laryngeal nerve, ITA – inferior thyroid artery, TG – thyroid gland, RCA – right common carotid artery, VAN – vagus nerve, BCT – brachiocephalic trunk

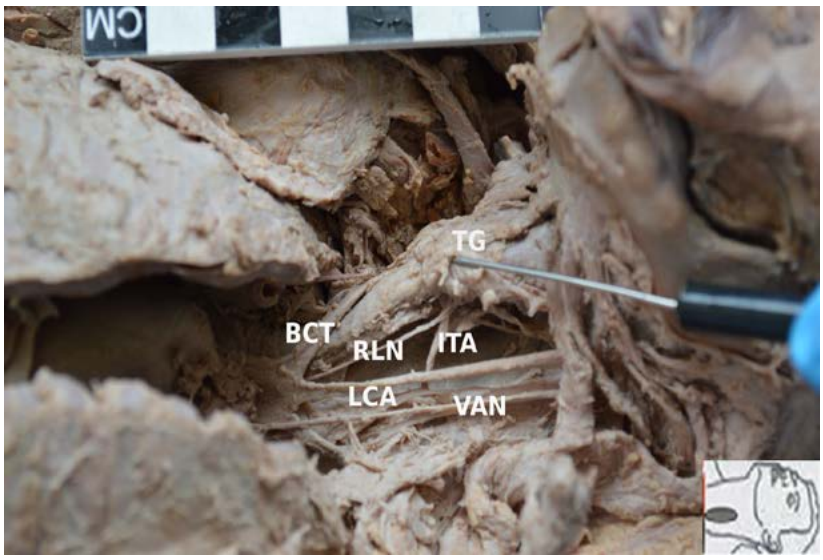

Figure 4: Photograph showing the RLN coursing anterior to the ITA on the left side

RLN – recurrent laryngeal nerve, ITA – inferior thyroid artery, TG – thyroid gland, LCA – left common carotid artery, VAN – vagus nerve, BCT – brachiocephalic trunk

of extra-laryngeal branches of the RLN which was observed was two. Branching of the RLN was more evident in term infants than in preterm infants.

Discussion

Thyroidectomy is the most performed endocrine procedure in surgery and its indications include benign and malignant conditions. Whereas more than 60% of thyroidectomies in adults are for benign diseases such as Graves' disease and colloid goitre**,** the main indication in children is papillary thyroid carcinoma.[11,12] Among the most feared complications following thyroidectomy is damage to the RLN, which is significantly higher in paediatric patients than it is in adults.[1] The main aim of this study was thus to document the anatomy of the RLN in paediatric cadavers, including the variations of its anatomical relations in the neck. Key findings from the study show that a non-recurrent RLN is uncommon and the relationship between the RLN and the TEG is more intimate on the left side of the neck. Furthermore, more than half of RLNs in children, irrespective of laterality, pass anterior to the ITA and extra-laryngeal branching of the RLN was seen in close to a third of cases.

The purported reasons for the higher rate of RLN damage during thyroidectomy in children include limited experience among surgeons and the complexity of the procedure as children with thyroid cancer commonly present at an advanced stage necessitating concomitant cervical lymph node dissection.[12] Another factor which puts the RLN at risk of injury is its anatomical course, which is known to be highly variable and unpredictable.[9]

The possible causes of injury to the RLN during thyroidectomy include accidental transection, stretching, thermal injury, suctioning, ischaemia or desiccation.[5] Early identification and tracking of the RLN during thyroidectomy has been shown to be among the critical measures to reduce the incidence of its injury. The inferior, medial, and lateral approaches for identification of the RLN, are based on the preference of the surgeon. The landmarks which are frequently used to identify the RLN include the thymus lower down in the neck, the TEG, ITA and the ligament of Berry, but none have been found to be reliable.[13]

In the current study, over 96% of the left RLNs ran within 0–15 degrees to the TEG as compared to 65% of the nerves on the right side. Around 66% of RLNs in the current study travelled close to the TEG, which is within the range (24.9–100%) reported by Henry et al.[14] The majority of RLNs that followed the course of the TEG were found on the left side (88.9%) which is contrary to < 70%, which was reported by Henry et al. in 2017.[9] Symmetry of the anatomical relationship of the RLN and the TEG was observed in 40.7% of the cadavers

in the current study, which is less than the pooled prevalence of 61.1% reported by Henry and colleagues.[9] This finding cautions that the TEG is a less reliable landmark for initial identification of the RLN during thyroidectomy in children as compared to a landmark such as the ligament of Berry.[9]

The recurrent laryngeal nerve either passes posterior or anterior to the ITA, or between its branches. Findings from the current study showed that on the right side the RLN coursed predominately anterior to the ITA (68%) and less so between its branches (13%). Comparatively, the RLN coursed anterior to ITA on the left in 57% and between its branches in 3.3% of the cadavers. The predominance of the anterior course of the right RLN in relation to the ITA mirrors findings by Henry and colleagues.[14] The rarity of the RLN traversing between the branches of the ITA is in keeping with findings from all prior studies.[15,16]

In the current study, the rate at which the relationship of the RLN and ITA was replicated bilaterally was less than 50%. In a meta-analysis of 79 studies, a symmetrical relationship between the RLN and ITA was observed in 36.6% of the cases. Henry et al.[14] found that 50.7% of all RLNs assume a posterior course relative to the ITA. In addition, Ling et al.,[15] analysing 32 studies with a total of 8 655 nerves found that 51.0% of all RLNs will course posterior to the ITA. The RLN and ITA therefore have a variable anatomy which all surgeons need to be aware of to avoid complications during thyroidectomy. A combination of miniature size and limited experience of a surgeon can make thyroidectomy exceedingly difficult in children, thus elevating the risk of injury to the recurrent laryngeal nerves.

Close to a third (29%) of RLNs in this study demonstrated extra-laryngeal branching. In a meta-analysis of 69 studies comprising 28 387 RLNs, Henry et al. demonstrated that 60% of RLNs gave extra-laryngeal branches.[14] The finding of fewer extra-laryngeal branching in the current study might have been due a smaller sample size as it only included 61 RLNs. However, all the studies cited by Henry and colleagues were done in adults. It is possible that some of the extra-laryngeal branches of the RLN in the study were small and could not be identified. The mean diameter of the left RLNs from the current study was 0.74 ± 0.28 mm as compared to the median size of 1.51 mm reported by Saito et al. from measurements conducted during thoracoscopic oesophagectomy in adult patients.[18] Thomas et al.[19] reported more extra-laryngeal branching of the RLN on the right side than on the left side. In the same study, symmetrical bilateral bifurcation of the RLN was observed in 16.4% of the dissections. A wide range of extra-laryngeal branching ranging from 5 to 100% with a symmetrical (bilateral) branching of 36.5% was reported in a systematic review of 28 387 RLN by Henry and colleagues in 2016.[14] The same authors reported bifurcation, trifurcation and more than three branches of RLN in 51.1%, 4.7% and 2.2%, respectively.[14] Prades and colleagues, however, reported even higher rates of extra-laryngeal branching of the RLN including bifurcation (88%) and trifurcation (12%) but in only 0.2% of cases were more than three branches present.[20]

Strength and limitations of the study

The intended stratification for age and gender was not feasible due to the limited number of specimens. Despite prior dissection of these cadavers, the neck anatomy was not distorted therefore the recorded findings are reliable. Notwithstanding the relatively small number of cadavers, the findings were significant.

Conclusion

In our study of paediatric cadavers, the left RLN ran within 0–15º of the TEG in 96% of the cases. Around 68% of right RLNs passed anterior to the ITA. Close to 30% of the RLNs gave branches. Symmetry of anatomy of RLNs was found in less than 25% of cases. The anatomy of the RLN in children is highly variable, and therefore, findings on the contralateral side should not be relied on to guide dissection on the opposite side during thyroidectomy in children.

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Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

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Paediatric liver injury – an eight-year retrospective review from a major trauma centre in South Africa

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Background: We benchmark our institutional experience with paediatric liver trauma in a low- to middle-income country (LMIC) to identify areas pertinent to its successful management.
Methods: Retrospective review of characteristics and outcomes of children less than 18 years old with liver trauma between December 2012 and October 2020 at a major trauma centre in South Africa.
Results: Seventy-five children sustained liver trauma; 53 (71%) male and 22 (29%) female. Events were more frequent in rural areas (60%), which had significantly longer time to hospital admission than urban areas (mean 10 vs 7.3 hours). There were eight (10%) penetrating mechanisms of injury. On presentation, mean shock index (SI) was 1.04 (SD 0.28) and median revised trauma score (RTS) was 7.84 (range 4–7.84). Children aged 0–5 years had significantly higher SI (mean 1.3 vs 0.88) and lower RTS than the 16–18 age group (median 7.2 vs 7.8). The mean injury severity score (ISS) was 19 (SD 9.24). The median American Association for the Surgery of Trauma (AAST) grade of liver injury was 2 (range 1–5). Twenty-three (31%) children underwent a laparotomy; six (8%) of which received immediate damage control surgery (DCS). Twenty-eight (37%) children developed one or more complications during admission. Forty-seven (63%) children required intensive care unit (ICU) admission, of which 29 (38.7%) required ventilation. Three (4%) children died; two having undergone a laparotomy.
Conclusion: Selective non-operative management and DCS principles have achieved low mortality rates in children with liver trauma in our centre. However, significant ICU and respiratory resources are still needed to manage the associated morbidity.
Keywords: liver trauma, abdominal injury, paediatric, damage control surgery, selective non-operative management, trauma centre, global surgery

Introduction

There has been a radical change in the management of paediatric intra-abdominal solid organ injury over the last fifty years from an operative approach to a selective non-operative management (SNOM) strategy.[1-5] This transition has been supported by the ongoing improvements in imaging, as well as by advances in interventional radiology and critical care. SNOM has been widely applied to liver trauma, with impressive results.[5-7] Furthermore, when surgery is required, there has been a move away from major liver procedures towards liver packing. These packs are left in situ to control bleeding until the patient's physiology has been restored. Once this has been achieved, the patient is returned to theatre and the packs are removed. Liver trauma is the prototypical example of the now widely accepted concept of damage control surgery (DCS).[4,5] The principles of DCS have, in turn, been extended to other injuries and to other areas such as damage control anaesthesia (DCA) and resuscitation.

Recently, there has been increased concern and awareness surrounding discrepancies in healthcare access and surgical outcomes between different population groups and regions. The Lancet Commission on Global Surgery (LCOGS) has highlighted that the vast majority of surgical procedures are performed in high-income countries (HIC) despite the overwhelming burden of surgical pathology being in low- and middle-income countries (LMICs).[8,9] In light of this, the LCOGS has encouraged scholars to explore differences in the care of common surgical conditions in order to quantify discrepancies in access, management and outcomes according to the income level of different countries. If access to care is hindered, then outcomes will be less than ideal. In addition, failure to apply modern and contemporary concepts such as SNOM and DCS may also impact outcomes. For these reasons, we aim to provide a benchmark of our experience with paediatric liver trauma to identify potential discrepancies in access and outcomes of surgical care compared to international experience.[10-12]

Materials and methods

Grey's Hospital is a tertiary care public hospital located in Pietermaritzburg, the capital of the KwaZulu-Natal (KZN) province in South Africa. This province on the eastern seaboard of South Africa is the most populous province in the country. It is beset by huge discrepancies in wealth and many of its regions score very poorly in terms of quality of life and poverty indicators. The Pietermaritzburg Metropolitan Trauma Service (PMTS) covers a catchment of over 3 million people. Nineteen other rural hospitals within the province refer to PMTS. Annual admissions exceed 4 000, over 50% of which are penetrating injuries. A prospectively entered surgical registry known as the hybrid electronic medical registry (HEMR) has been maintained by the PMTS since 2011. This system captures data on all admissions to our trauma centre.

Weekly morbidity and mortality meetings are held, and stringent management protocols are followed to deliver optimal and universal care. All presenting patients are resuscitated according to advanced trauma life support principles. DCS involved immediate exploratory laparotomy. This occurred in patients who presented haemodynamically unstable, peritonitic, or with eviscerated hollow organs or with pneumoperitoneum. In other patients, SNOM is followed. This involves keeping the patient fasted, with intravenous fluids and analgesia and regular monitoring for the next 12 hours. If they remain stable, they are fed and observed for further 12–24 hours. Patients may be discharged following this observation period or transferred back to domicile hospitals for further observation at the discretion of the treating surgeon. Failure of SNOM is defined as delayed laparotomy (DELAP). Other procedures unrelated to the abdomen, such as fracture fixations or neurosurgical interventions, were considered as SNOM.

Children aged less than or equal to 18 years old with liver injuries admitted to our centre between December 2012 and October 2020 were identified from the HEMR database. Patient demographics, details of injury (time, location, mechanism), admission vital signs and biochemistry, management, complications, length of stay (LOS) and clinical outcomes were analysed.

Shock index (SI) is calculated as heart rate divided by systolic blood pressure; a higher score is associated with degree of shock. The revised trauma score (RTS) quantifies severity of trauma injuries based on Glasgow Coma Scale (GCS), blood pressure and respiratory rate; a higher score is associated with higher survival. The injury severity score (ISS) standardises the severity of traumatic injury based on the worst injury of six body systems; a higher score is associated with greater mortality.

Statistical analysis

Relevant data were extracted to Excel. Statistical analysis was performed using R (version 4.0.2; R Foundation for Statistical Computing, Vienna, Austria). Parametricity was assessed via the Shapiro–Wilk test. Continuous variables with normal and non-normal distributions were compared using independent samples t-test and Mann–Whitney U test, respectively; then reported using mean with standard deviation (SD) and median with range, respectively. Categorical variables were compared using the chi-squared test with the Fisher's exact test used when one group had less than five events. Statistical significance is achieved when $p < 0.05$.

Results

Over the eight-year period (December 2012 to September 2020), 75 children were identified with a traumatic liver injury. The mean incidence was eight (SD 3.5) cases per annum. There were 22 girls and 53 boys with a median age of 10 (range 1–18). Time to hospital admission was 7.3 (SD 6, $n = 18$) and 10 (SD 4.9, $n = 26$) hours from time of injury in urban ($n = 30$, 40%) and rural ($n = 45$, 60%) locations, respectively ($p = 0.04$). There were eight (10%) penetrating

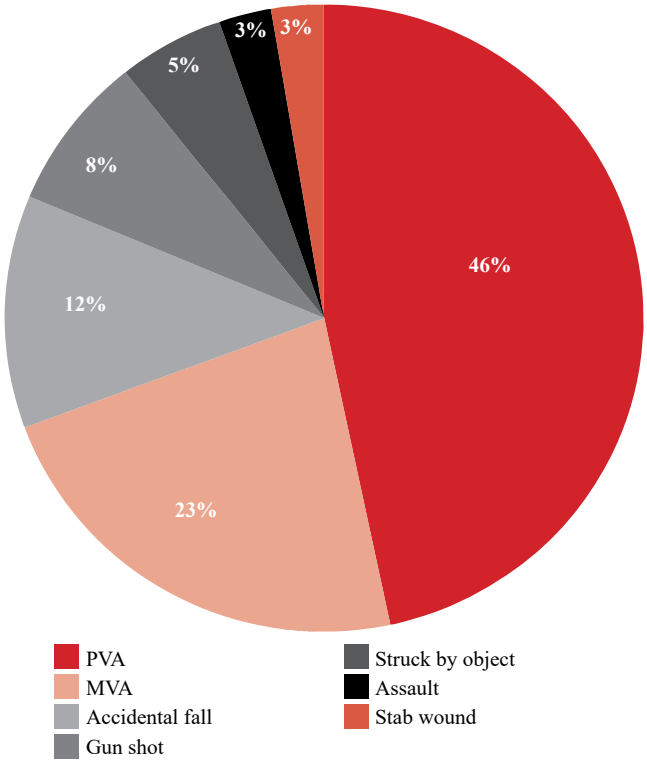

Figure 1: Mechanism of injury to children with liver trauma, n = 75

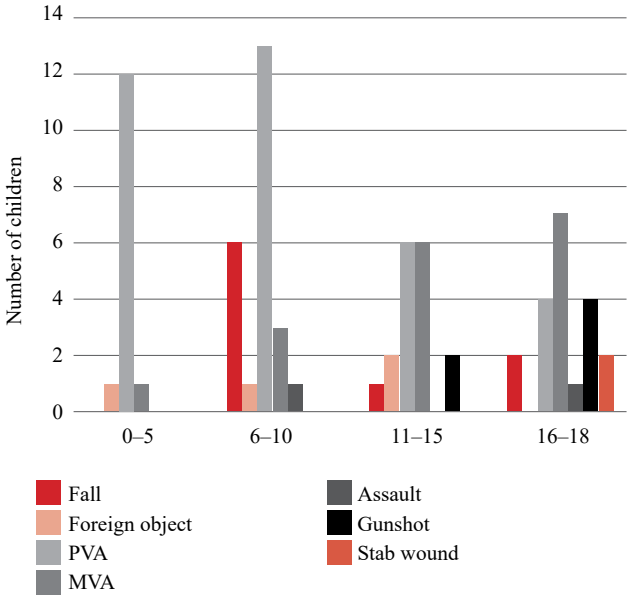

Figure 2: Mechanism of injury to children with liver trauma according to age, n = 75

mechanisms, of which six were gunshot wounds (GSWs) and two stab wounds. Of the 67 (90%) blunt mechanisms there were 35 pedestrian vehicle accidents (PVA), 17 motor vehicle accidents (MVA), nine falls, four impact from foreign object and two assaults. Penetrating wounds and MVA were more frequent in older children ($p = 0.008$). Mechanism of injury by cohort and age is shown in Figure 1 and Figure 2, respectively.

Physiology

On presentation, mean SI was 1.04 (SD 0.28, $n = 63$) and mean RTS was 7.84 (range 4–7.84, $n = 61$). Children aged 0–5 years had significantly higher SI than those aged 6–10, 10–15 and 16–18 (mean 1.3, 1.06, 0.9 and 0.88, $p < 0.001$, $n = 70$). Children in the 16–18 age group had significantly higher RTS scores compared to 0–5 and 6–10 age group (median 7.84 vs 7.2 and 7.6, $p = 0.04$, $n = 61$). Admission biochemistry included median arterial pH 7.36 (range 6.95–7.53, $n = 63$), BE -3.35 (-19.5–15.8, $n = 62$) mmol/L, lactate 1.65 (-1.7–12.4, $n = 53$) mmol/L, Hb 10.9 (6.2–20.1, $n = 61$) g/dl. The mean ISS was 19 (SD 9.24, $n = 61$). Table I summarises patient characteristics and management by year. There was an increasing temporal trend for older children to sustain liver trauma (median 4 to 13 years old in 2013 and 2019, respectively). No statistical differences in IS, RTS or ISS were noted between rural and urban areas of trauma or between blunt versus penetrating mechanisms of injury.

Imaging

Sixty-seven (89.3%) children underwent a CT scan. Eight children proceeded directly to DCS without CT. Children involved in PVA were significantly more likely to receive a CT scan (100%) and those involved in GSW or stab wounds were more likely to forego a CT scan ($p < 0.05$). Liver trauma in children without imaging was identified intraoperatively. No statistically significant differences in SI, RTS and ISS were observed between children who received CT and those who did not.

Grade of liver injury

The median grade of liver injury was 2 (range 1–5). Grade of liver injuries based on imaging included 16 patients (24%) with grade I, 21 (28%) with grade II, 19 (25.3%) with grade III, 10 (13%) with grade IV, and five (6.7%) with grade V injuries. Liver injury was recorded in five patients intraoperatively: one grade I, two grade II, one grade IV and one grade V. In two patients, the grade was not recorded. The American Association for the Surgery of Trauma (AAST) for the liver showed a positive trend for ISS score (25 vs 18 between grade I and V) though no associated trends in management or outcomes were observed (Figure 3).

Concurrent injuries

Four children sustained an isolated liver injury whilst the majority (95%) sustained injuries to multiple structures and organs including: chest in 42 (56%), abdomen in 40 (53%), fractures to extremities and pelvic girdle in 34 (45%), head or spinal injuries in 40 (53%) and face in 10 (13%) children. The most commonly injured organs associated with liver injury included: 16 (21.3%) renal injuries, 11 (14.7%) splenic, 11 (14.6%) enteric, eight (10.7%) urogenital, seven (9%) adrenal, three (4%) pancreas, two (3%) diaphragm and one (1%) gastric injury. Concurrent intrabdominal vascular

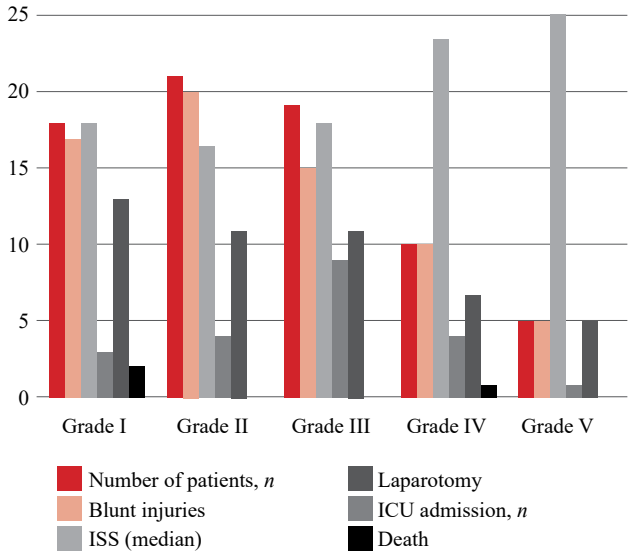

Figure 3: Characteristics and management of children with liver trauma according to liver AAST, n = 75

Table I: Characteristics, management and outcome of children with liver trauma by year

	2012	2013	2014	2015	2016	2017	2018	2019	2020
Number of patients, *n*	2	5	12	11	10	13	7	8	7
Age, median	16.5	4	7	9	10	13	11	14	13
ISS, median	21.5	20	14.5	17	22.5	17	18	22	13
RTS, median	6.85	7.2	7.8	7.8	7.55	7.84	6.9	7.69	7.8
Liver AAST, median	2.5	3	2	2	3	3	2	3	2
Whole body CT scan	1	2	7	6	7	9	4	7	3
SNOM	2	4	10	11	9	13	6	8	6
DCS	0	1	2	0	1	0	1	0	1
DELAP	1	1	3	5	1	3	1	2	0
Total laparotomies	1	2	5	5	2	3	2	2	1
ICU admission, *n*	1	5	9	9	6	4	5	6	2
LOS, median days	11	10	6.5	7	4	7	12	14.5	1
Deaths, *n*	0	1	0	0	1	0	0	0	1

ISS – injury severity score, RTS – revised trauma score, AAST – American Association for the Surgery of Trauma, SNOM – selective non-operative management, DCS – damage control surgery, DELAP – delayed laparotomy, LOS – length of stay

Table II: Characteristics and management of children with liver trauma receiving laparotomy versus no laparotomy

	Laparotomy	No laparotomy	*p*-value
Number of patients, *n*	23	52	
Age, median (IQR)	14.45 (8.32, 17.02)	9.76 (5.44, 14.55)	0.051
Gender (%)			
Male	15 (65.2)	38 (73.1)	0.585
Female	8 (34.8)	14 (26.9)	
Mechanism (%)			
Blunt	16 (69.6)	51 (98)	0.0008*
Penetrating	7 (30.4)	1 (1.9)	
SI, median (IQR)	1 (0.86, 1.15)	1.01 (0.83, 1.24)	0.479
RTS, median (IQR)	7.84 (7.84, 7.84)	7.55 (6.90, 7.84)	0.007*
ISS, median (IQR)	18.00 (16.00, 25.00)	18.00 (13.00, 25.50)	0.994
Liver AAST (%)			
I	3 (14.3)	15 (28.8)	0.192
II	4 (19)	17 (32.7)	
III	9 (42.9)	10 (19.2)	
IV	4 (19)	6 (11.5)	
V	1 (4.8)	4 (7.7)	
WBCT (%)	6 (26.1)	40 (76.9)	< 0.001*
ICU admission (%)	13 (56.5)	28 (53.8)	1
Ventilation (%)	7 (30.4)	22 (42.3)	0.474
LOS, median (IQR)	10.00 (4.50, 16.00)	5.00 (1.00, 11.25)	0.033*
Deceased (%)	2 (8.7)	1 (1.9)	0.22

RTS – revised trauma score, ISS – injury severity score, AAST – American Association for the Surgery of Trauma, WBCT – whole body CT, LOS – length of stay
*Statistically significant, $p < 0.05$

injury was noted in four (5%) children. These injuries provided a median ISS of 18 (range 4–43, $n = 61$). ISS by age was not significantly different ($p = 0.98$, $n = 61$).

Management

Twenty-nine children required intubation; 11 were intubated at the scene or after arrival at hospital trauma bay. Twenty-three children required laparotomy (31%); six of which were damage control procedures. Sixty-nine children were managed with SNOM; 17 (24.6%) failed and received DELAP. The mean laparotomy rate was found to be 30.4% (SD 13.8) per year with no statistically significant temporal trends ($p = 0.70$). Procedures performed at laparotomy included liver packing (6), hepatic drainage (6), intestinal injury repair (5), splenectomy (3), nephrectomy (3), washout only (2), pancreas debridement (1), loop ileostomy (1), bowel resection (1), gastric perforation repair (1), aortic graft patch (1), inferior vena cava (IVC) repair (1) and laparostomy closure (2). Ten fractures required intra-operative fixation: three via open reduction and internal fixation (ORIF), two via nail, one via external fixation, one via screw, one via k-wire and one mandible stabilisation. One child required a suprapubic catheter following a vaginal laceration.

Children who underwent a laparotomy had significantly higher RTS (median 7.84 vs 7.55, $p = 0.007$) sustained significantly more penetrating injuries (30.4% vs 1.9%) and had significantly longer LOS (median 10 vs 5 days, $p = 0.03$) than children who were managed non-operatively (Table II). Greater SI scores were observed in children who received DCS versus SNOM; median 1.14 (1–1.25) vs 0.89 (0.59–1.39) ($p < 0.05$). No children received exploratory laparoscopy.

Table III: Complications of children with liver trauma receiving laparotomy versus no laparotomy

	Laparotomy	No laparotomy	*p*-value
Number of patients, *n*	23	52	
Any complication, %	12 (52.2)	16 (30.8)	0.131
Relook laparotomy, %	6 (26.1)	0 (0)	0.0001*
Abdominal, %	5 (21.7)	1 (1.9)	0.014
Wound, %	4 (17.4)	1 (1.9)	0.029*
Respiratory, %	6 (26.1)	14 (26.9)	1
Neurological, %	4 (17.4)	7 (13.5)	0.728
Renal, %	4 (17.4)	0 (0)	0.007*
Haematological, %	2 (8.7)	1 (1.9)	0.221
Deaths, %	2 (8.7)	1 (1.9)	0.221

*Statistically significant, $p < 0.05$

Outcomes

Twenty-eight (37%) children developed one or more complications during admission. Twenty (27%) children developed a respiratory complication: ten required antibiotics for pneumonia, while three required tube thoracostomy to manage parapneumonic effusion. One child experienced repeated failed extubation secondary to subglottic stenosis requiring endoscopic dilatation. Six children required tracheostomy. Six children (21%) required repeat laparotomy (three for removal of packs, one for a washout,

Table IV: Characteristics and management of children with liver trauma comparing survivors to non-survivors

	Survivor	Non-survivor	*p*-value
Number of patients, *n*	72	3	
Age, median (IQR)	9.90 (6.10, 15.84)	14.63 (14.54, 14.82)	0.251
Gender (%)			
Male	51 (70.8)	2 (66.7)	1
Female	21 (29.2)	1 (33.3)	
Mechanism (%)			
Blunt	65 (90.2)	2 (66.7)	0.29
Penetrating	7 (9.7)	1 (33.3)	
RTS, median (IQR)	7.84 (6.90, 7.84)	5.97 (5.03, 6.90)	0.507
ISS, median (IQR)	18.00 (13.25, 25.00)	37.00 (23.00, 40.00)	0.262
Liver AAST (%)			
I	16 (22.9)	2 (66.7)	0.223
II	21 (30)	0 (0)	
III	19 (27.1)	0 (0)	
IV	9 (12.9)	1 (33.3)	
V	5 (7.1)	0 (0)	
WBCT (%)	45 (62.5)	1 (33.3)	0.555
SNOM (%)	51 (70.8)	1 (33.3)	0.221
Laparotomy (%)	21 (29.2)	2 (66.7)	0.221
ICU admission (%)	38 (52.8)	3 (100)	0.246
LOS, median (IQR)	7.00 (2.00, 14.00)	1.00 (1.00, 3.00)	0.147

RTS – revised trauma score, ISS – injury severity score, AAST – American Association for the Surgery of Trauma, WBCT – whole body CT, SNOM – selective non-operative management, LOS – length of stay

one for ischaemic gut and one for infected mesh removal). Three children required multiple relook laparotomies (two for further washouts and one for an intra-abdominal bleeding diathesis). Six (21%) children developed abdominal complications which included postoperative collections, biloma, volvulus, postoperative ileus and iatrogenic uterine perforation. Four (14%) children developed wound complications which included postoperative sepsis, mesh infection, pressure-induced decubitus ulceration, and digital ischaemia secondary to a peripheral arterial line. Eleven (39%) children developed neurological complications which included seizures, delirium, post-traumatic stress disorder, depression, neuroleptic malignant syndrome, post external ventricular drain (EVD) related brain injury, and three children required repeat CT brain for change in GCS. Three (11%) children developed renal complications with one child requiring renal replacement therapy. Three (11%) children developed haematological complications including postoperative deep vein thrombosis, coagulopathy and thrombocytosis.

Children who underwent a laparotomy had significantly more abdominal (5 vs 1, $p = 0.01$), wound (4 vs 1, $p = 0.03$) and renal (4 vs 0, $p = 0.007$) complications than children who did not (Table III).

The average LOS was a median 7 (range 1–43) days. Children who received laparotomy had median LOS of 10 days vs 5 days for those without laparotomy ($p = 0.03$, Table II). A total of 47 (62.6%) children required admission to the intensive care unit (ICU) with a median LOS of 4 (range 1–42, $n = 41$) days. Twenty-nine (38.7%) children required mechanical ventilation and four (5.3%) required inotropic support.

A higher RTS score was observed in children without ICU admission and ventilatory support; median 7.84 (range 6.38–1.47) vs 7.55 (4–7.8) and median 7.8 (7.55–7.8) vs 6.9 (4–7.8), respectively ($p < 0.05$). A higher ISS score was observed in children who required ICU admission; median 20 (4–39) vs 16 (5–43) ($p < 0.05$).

Mortality

Three (4%) children did not survive, two of whom had undergone a laparotomy. Two of the deaths were secondary to hypovolaemic shock; one following PVA and another secondary to GSW. One death was due to ischaemic bowel secondary to blunt trauma. The children that died were older (median 15 vs 10, $p = 0.3$) and had higher ISS (37 vs 18, $p = 0.26$) than survivors (Table IV).

Discussion

Paediatric liver trauma has a different profile from its adult equivalent. With the exception of a defined subset of patients who sustain penetrating trauma, the mechanism of injury is predominantly blunt in nature.[7] Blunt liver trauma is the prototypical indication for both SNOM and DCS.[4-6] The widespread availability of accurate CT assessment and grading of liver injuries following blunt abdominal trauma has facilitated SNOM. Just under 90% of patients in this series underwent a CT scan, and 69% of children were managed non-operatively.

Our operative experience reflects the trend towards a minimalistic approach away from major liver resections; no liver resections were performed in our cohort. The most common operative strategy was liver packing followed by hepatic drainage, which is in keeping with the trend towards surgical minimalism seen in the literature. Furthermore, DCS

was undertaken in just under one-third of surgical cases. This series differs from most reported in HICs in that 10% of the patients sustained penetrating trauma. Penetrating trauma is not analogous to blunt trauma and different management strategies and prerogatives apply.[13] We observed a higher rate of penetrating injuries in the operative compared to the non-operative group (30.4% vs 1.9%). Most series of paediatric liver trauma from international centres in HICs have almost no penetrating mechanisms and this likely explains their higher overall rate of non-operative management.[10-12] Furthermore, a pattern of injury and management profile approaching that of an adult population was observed in children receiving laparotomy which may be influenced by the older median age of this group.

The management of paediatric liver trauma requires substantial resources as evidenced by the significant associated morbidity, where over one-third of children developed a complication and almost two-thirds required ICU admission. Although SNOM significantly reduced abdominal-, renal- and wound-related complications, respiratory complications are still frequently encountered and require significant resources in terms of ICU and pulmonary support.[10-12]

Legal versus biological definitions of children vary and some countries use the age of puberty to highlight the physiological differences in children and to assist allocation to paediatric facilities. Children have different compensatory mechanisms and communication barriers compared to adults. Huang et al. observed a mean ISS score above 5 and RTS score below 7.67 was found in children admitted to ICU, comparable to our mean scores of 21 and 7, respectively.[14] An SI > 1.0 has been widely found to predict increased risk of mortality and other markers of morbidity; in our centre an SI of median 1.14 was observed in children receiving DCS.[15] Clinical implementation of trauma scoring systems could be investigated to provide objective and accessible measures to facilitate intervention.

The mortality rate for liver trauma in children in our centre is less than five per cent which is in keeping with most international HIC centres.[10-12] This suggests that we are applying the modern principles of SNOM and DCS appropriately. Reducing this mortality further will be challenging since it is likely an outcome of the cumulative severity of the trauma, as illustrated by the high ISS scores in this series, rather than the severity the liver injury.

Conclusion

Liver trauma in children is still associated with a high morbidity rate, although there have been dramatic improvements in mortality over the last three decades. SNOM and DCS principles have been applied to children with trauma in our centre; however, the successful management of liver trauma requires access to significant ICU resources in this LMIC.

Conflict of interest

The authors declare no conflicts of interest.

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Ethical approval

Ethical approval for the hybrid electronic medical registry has been granted by the Biomedical Research Ethics Committee of the University of KwaZulu-Natal (ref. no BE207/09 BCA 221/13).

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