Endoscopic surgery in children: Better understanding of pathology

9 September 2006, SAAPS Congress, Sun City

KLASS (N.) M. A. BAX, M.D., PH.D., F.R.C.S. (ED.)
Professor of Paediatric Surgery and Head of Department of Pediatric Surgery, Erasmus Medical Center – Sophia Children’s Hospital, Erasmus University, Rotterdam, The Netherlands

It is a great honour to be invited to give the 2006 J. H. Louw Memorial Lecture. Professor Louw was a great man. His interest in paediatric surgery was undoubtedly stimulated by his meeting with David Waterstone from the Hospital for Sick Children in London during World War II and the fact that his firstborn son died of intestinal atresia. In 1951 he did a fellowship under Sir Dennis Brown at the Hospital for Sick Children and there started his work on the pathogenesis of intestinal atresia.

Professor Louw is without doubt the father of paediatric surgery in South Africa. In 1952 he established the first paediatric surgical service in Cape Town at Groote Schuur Hospital. He was one of the promoters of Red Cross War Memorial Children’s Hospital, and set up a training programme in paediatric surgery there. His prerequisite for any surgeon working at Red Cross Hospital was that the surgeon would be capable of looking after the child as a whole, and I feel that this should still hold true today. He was the first president of the South African Association of Pediatric Surgeons, a position he held from 1976 until 1980. In 1980 he received the Dennis Brown Medal, the highest award of the British Association of Paediatric Surgeons.

Professor Louw was not only a paediatric but also a general surgeon. He was head of the Division of Surgery of the University of Cape Town in 1955 - 1981. I worked in Cape Town in 1974 - 1977 at both Groote Schuur and Red Cross hospitals, during which time I was his personal registrar for 12 months, so I came to know him well.

Like Lord Moynihan, Professor Louw believed that surgery is an instrument of research by lifelong direct observation of the pathology of the living. His work on the pathogenesis of intestinal atresia is a good example of this belief. He went from surgical observation to the laboratory, and together with Chris Barnard published the classic paper on the vascular pathogenesis of intestinal atresia in The Lancet in 1955.

Endoscopic surgery in children

At Wilhelmina Children’s Hospital in Utrecht we have been doing pioneering work in the field of endoscopic surgery in children since 1992. Since then we have performed more than 2 000 endoscopic surgical procedures. One-third were in children less than 6 months of age and more than 85% were therapeutic.

Two major aspects of endoscopic surgery stand out: on the one hand minimal access, resulting in less pain, quicker recovery, a shorter hospital stay and better cosmetic outcome, and on the other hand the degree of magnification. When you ask people how much magnification is obtained, nobody knows. This is not surprising, as many variables determine magnification, such as telescope brand, angle and diameter, distance between the telescope and the region of interest, degree of optical zoom, screen diameter, and the surgeon's distance from the screen. Together with Karl Storz, of Tutlingen, Germany, we looked at magnification in a particular setup: a specific telescope and camera, and a 19-inch cathode array screen. Only 0° telescopes were used. The distance from the tip of the telescope to the target was set at 1, 2 and 3 cm. The optical zoom was set once at minimum magnification and once at maximum magnification. Using a 5 mm telescope at 1 cm distance from the target, magnification was about 10 times with minimal zoom and about 40 times with maximal zoom. At 2 cm distance, magnification was about half and at 3 cm about one-quarter.

Yet the better view obtained with endoscopic surgery is not only due to magnification. In contrast to open surgery, the viewing field in endoscopic surgery is free from gauzes, fingers or retractors. Besides, thoracoscopic or laparoscopy will hardly disturb the initial anatomy and pathology.

Endoscopic surgery: Better understanding of pathology

What is more, endoscopic surgery has not only led to better surgery because of a more detailed view and less extensive dissection, it has also led to better understanding of pathology, which I shall demonstrate with a number of examples.

Oesophageal atresia

Azygos lung lobe. In thoracoscopic correction of oesophageal atresia we made several interesting observations. The first example is an azygos lung lobe (Fig. 1). Although this lobe does not need treatment, the sac is in the way when repairing oesophageal atresia. Just making a hole in the sac is likely to result in herniation of the lung through this opening. In order to avoid such a complication, the azygos vein should...
be transected and the sac incised. An azygos lobe is seen in 0.5 - 1% of anatomical dissections and routine chest radiographs.\textsuperscript{5,6}

Relative position of the distal fistula to the azygos vein. We also learned that in at least 30% of cases of oesophageal atresia the distal fistula is located above the azygos vein (Fig. 2). In these patients there is no need to sacrifice this vein.

Proximal oesophageal innervation from the recurrent laryngeal nerves. In a patient with membranous-type oesophageal atresia with distal fistula, the upper oesophagus had to be mobilised circumferentially in order to isolate the distal fistula. In that case we could identify the left recurrent laryngeal nerve from behind. Moreover, the left recurrent laryngeal nerve clearly supplied branches to the upper oesophagus (Fig. 3). This is in line with observations made by Davies in postmortem dissections of children with oesophageal atresia. Davies found that the recurrent laryngeal nerves supply motor branches to the upper oesophageal pouch.\textsuperscript{7} This brings into question the safety of extensive mobilisation of the upper oesophagus in oesophageal atresia. Moreover, in a patient with oesophageal atresia with distal fistula, I estimated the gain in length before and after mobilisation of the proximal pouch and came to the conclusion that mobilisation does not add much to length. One can argue that such a dissection is needed in order to rule out a proximal fistula. The incidence of a double fistula is around 1%, which means that 99 dissections have to be carried out in order to find one proximal fistula.\textsuperscript{8} To my mind this is not worth while. Common sense also dictates that the distal oesophagus should not be mobilised, since this will lead to denervation and further impairment of the motor function of the oesophagus.

Relationship of the phrenic nerve to the superior caval vein. Thoracoscopy in a child in need of an aortopexy for tracheomalacia showed a persistent left superior caval vein with the phrenic nerve on top of it (Fig. 4). The prevalence of a left caval vein in children with oesophageal atresia is around 10%.\textsuperscript{9} The close proximity of the phrenic nerve to the caval vein on the right explains why the phrenic nerve is at risk during ablation for atrial fibrillation.\textsuperscript{10}

Pain in the slipping rib syndrome
In a patient with slipping rib syndrome, the tip of the 8th rib dislocated from the 7th rib on the right when the patient moved from a recumbent into a sitting position. This caused severe pain radiating to the back. We approached the region thoracoscopically and detached the diaphragm at that site. It could be seen clearly that the tip of the 7th rib dislocated from the 8th rib and that the 7th intercostal nerve was being pinched in between (Fig. 5). Impingement of intercostal nerves in the slipping rib syndrome has been implicated as a cause of the associated pain.\textsuperscript{11,12}

Identification of focal hyperinsulinism in the newborn
It has always been difficult to distinguish between diffuse and focal forms of congenital hyperinsulinism preoperatively,
yet the treatment implications are great: focal lesions are curable. Classic imaging is not very helpful, and until recently cumbersome tests such as transhepatic portal venous imaging and sampling or intra-arterial calcium stimulation together with hepatic venous sampling were needed.\textsuperscript{13,14} In 2002, Fekete reported that with the help of a magnifying glasses she had been able to see focal lesions in 31 out of 45 cases. She also used transhepatic portal venous imaging and sampling as well as preoperative frozen section biopsy analysis.\textsuperscript{15} Using the laparoscope, which has a much higher magnification rate, we were able to identify focal lesions in two neonates with congenital hyperinsulinism (Fig. 6).\textsuperscript{16,17} A newly introduced technique, 18F-dehydroxyphenylalanine positron emission tomography (18F-DOPA PET) scanning, has shown promising results in identifying focal lesions preoperatively.\textsuperscript{18,19} The next two of our patients with congenital hyperinsulinism had such a scan, which was negative in the first but positive in the second. In the first patient no focal lesion was found at surgery, and histological examination revealed diffuse disease in the laparoscopically resected tail. As could be expected, symptoms persisted. Laparoscopic subtotal spleen-sparing pancreatectomy was performed later on but was not curative either. In the patient with a positive 18F-DOPA PET scan the lesion was clearly seen but apparently insufficiently resected as symptoms persisted. At repeat endoscopic surgery a few weeks later the suspicious area was further resected together with the tail of the pancreas, resulting in cure. A preoperative 18F-DOPA PET scan in combination with laparoscopy seems the way to go in these patients. We also found that repeat laparoscopic surgery in these patients is much simpler than repeat open surgery, as there are far fewer adhesions than expected.

**Abnormal pelvic innervation in anorectal anomalies**

Several paediatric surgeons have found abnormal pelvic innervation in patients with anorectal anomalies. Classic studies are those of Scott in 1959 and of Stephens and Smith in 1971,\textsuperscript{20,21} and more recently Davies published on this issue.\textsuperscript{22} All concluded that the higher the anomaly, the more abnormal the innervation – in the sense that the pelvic splanchnic nerves run almost in the midline. Davies wrote that the hypogastric plexus is easily seen during surgery but that the pelvic plexus is usually not seen. In anorectal dysgenesis not only do the pelvic nerves run more medially than normal but Denonvilliers’ fascia is deficient, which explains why the surgeon may ‘get lost’ during posterior sagittal approaches. One of our patients with high-type anorectal anomaly had no functional urinary abnormalities before corrective surgery, and findings on preoperative miction cystourethrography were normal. Pre-pull-through distal colonography showed a high-ending blind rectum. At surgery a blind-ending rectum with fibrous connection to the base of the bladder was seen. It was almost impossible to enter the pelvis, even after dividing the fibrous cord close to the rectum. The hypogastric nerve plexus was clearly visible, but again it was almost impossible to enter the pelvis. The right kidney was located in the pelvis and both ureters ran on the sacrum close to the midline (Fig. 7). Eventually the rectum was pulled through on the right side. Unfortunately the patient had postoperative micturition...
problems, leaving little doubt that surgery interfered with innervation. Iatrogenic nerve damage may be difficult to avoid under certain circumstances. Laparoscopy helps in identifying abnormal innervation and may help in avoiding damage.

Bands of Ladd: do they really exist?
The bands of Ladd have puzzled me for a long time. To be honest, I never understood what they really were. Over and over again I read chapter V, on intestinal obstruction resulting from malrotation of the intestines and colon, in Ladd and Gross’s *Abdominal Surgery of Infancy and Childhood*, published in 1941. This chapter tells us that ‘improper rotation (which is almost always an incomplete rotation) produces characteristic findings of high intestinal obstruction resulting from external pressure on the second or third portions of the duodenum’, and ‘With incomplete rotation of the cecum one commonly finds this organ just below the distal half of the stomach and bands of reflected peritoneum running from it (or the ascending colon) to the right posterolateral abdominal wall. These bands or folds therefore lie directly across the descending portion of the duodenum and partially obstruct this viscus by external pressure. In a smaller number of cases the cecum has proceeded farther but is yet incompletely rotated, so that it lies directly over the duodenum and obstructs it by external pressure.’ Ladd and Gross then conclude: ‘It is obvious that there are really two separate lesions. One is concerned with the obstruction of the descending duodenum by an overlying cecum or peritoneal band. The other is concerned with the volvulus of the midgut.’

The pictures in this book are difficult to interpret. It looks as if the ileocolic region has twisted in a counterclockwise direction, while the small bowel has twisted in a clockwise direction. Yet it is stated that the volvulus in intestinal malrotation is always in a clockwise direction.

The true pathology of intestinal malrotation is difficult to unravel at laparotomy as it is usual to exteriorise the bowel first and then to untwist the bowel. In doing so the initial pathology is not appreciated. During laparoscopy, in contrast, the initial anatomy or pathological anatomy is hardly disturbed. We have laparoscopically explored 18 babies with symptomatic malrotation (i.e. presenting with bilious vomiting) within the first 6 weeks of life. All proved to have clockwise volvulus, but without necrosis at laparoscopy. We often noted distended veins in the mesentery or along the mesenteric bowel wall. One patient had chylous ascites and another had a fibrotic mesenterial stalk, indicating that the volvulus had been present for some time. In all it was clear that the obstruction was related to the volvulus and not to bands. It is the hepatic flexure of the colon rotating around the mesenteric stalk that drags the lateral peritoneal reflection of the ascending colon and hepatic flexure with it (Fig. 8). The so-called bands of Ladd are not odd embryonic structures, but just the peritoneal fixations of the hepatic flexure of the colon and adjacent ascending colon.

Conclusion
Mr Chairman, ladies and gentlemen, I would like to conclude – with Lord Moynihan and Professor Louw – that surgery
is an instrument of research and that the surgeon has a lifelong opportunity for research by direct observation of the living. When new observational tools like endoscopic surgery become available, we should take full advantage of them.

REFERENCES