Hirschsprung’s disease
Definition and pathophysiology
Hirschsprung’s disease (HD) is a congenital condition characterised by the absence of ganglion cells in the submucosal (Meissner’s) and myenteric (Auerbach’s) plexus [1]. Aganglionosis occurs due to the arrest of the craniocaudal migration of the neural crest cells (NCCs) within the gastrointestinal tract. The migration of NCCs takes an average of 7 weeks, commencing at week 5 and completed by week 12 of gestation [2]. Due to the failure of the migration process the distal rectum is always aganglionic with variable proximal bowel involvement. Ganglion cells help facilitate relaxation of the bowel; subsequently aganglionosis results in functional obstruction and dilation of the proximal ganglionic bowel. The transition zone (TZ) lies between the aganglionic and ganglionic bowel. Short-segment, ‘classic’ HD, is limited to the rectosigmoid and occurs in 80% of cases. Long-segment disease accounts for the remaining 20% of cases with aganglionosis extending proximal to rectosigmoid. Total colonic aganglionosis (TCA) accounts for 3-8% of cases and involvement of the small bowel is rare [1]. Mortality increases as the length of bowel involvement and has been reported as high as 66% in TCA patients by the age of 8 years [3].

Aetiology
It has been reported to vary across ethnic groups: 1.5 per 10000 in Caucasians, 2.1 African-Americans and 2.8 in Asian populations [4]. The male to female ratio is 4 to 1, however in long-segment disease male: female ratio is 1 to 1. The overall rate of familial recurrence was found to be 7.6% in a 2015 systemic review [5], previously quoted rates of 2.6-6% [6,7]. Familial recurrence rates were 20% in TCA [5,8]. The recurrence risk to siblings ranges from 1-33% depending on the sex and extent of aganglionosis. The lowest risk of 1% is for a female sibling of a male patient with short segment disease, whilst a male sibling of a female patient with long segment disease has the highest recurrence risk of 33% [8]. Genetic counselling should be offered for these families. HD is a multifactorial congenital condition with approximately 20% of infants having one or more associated abnormality. Downs syn-
drome is reported as the commonest associated chromosomal anomaly of 2-15% [9]. There are multiple associated syndromes which include neurocristopathy syndromes, for example: Shah- Waardenburg, Yemenite deaf-blind, Piebaldism, Multiple endocrine neoplasia type II and Riley Day syndrome; Goldberg-Shprintzen syndrome and Congenital central hypoventilation syndrome [6]. Eight key genes found to be involved in HD include proto-oncogene RET involved in normal enteric nervous system (ENS) differentiation and accounts for 35% of sporadic cases and 50% of familiar cases; glial cell line derived neurotrophic factor (GDNF) and neurturin (NTN), which control ENS differentiation; endothelin B receptor (EDNRB) and endothelin 3 (EDN3) account for 10% of cases and endothelin converting enzyme 1 (ECE1), SOX10, and SIP1 genes [1].

**Presentation**
The majority of patients are diagnosed with HD as a neonate. Patients generally present with failure to pass meconium within the first 24-48 hours of life, abdominal distension and bilious vomiting. If they present as older children symptoms can include chronic constipation, abdominal distension, vomiting, enterocolitis and failure to thrive, see Table I [1,10]. Enterocolitis is characterised by abdominal distension, offensive stool and sepsis, and has significant morbidity if left untreated. Downs syndrome patients are more susceptible to enterocolitis with an incidence as high as 50% and symptoms may become chronic [3,11,12]. This may then lead to faecal incontinence (at 12 years as high as 87%) [3]. A detailed history and examination is performed to ascertain key symptoms and signs. Rectal examination typically shows an explosive decompression of flatus and stool. Differential diagnosis is shown in Table II.

**Investigations**
A plain abdominal x-ray classically shows distended loops of bowel with paucity of gas within the pelvis (Fig. 1). The benefit of performing a subsequent contrast enema is to help locate the transition zone to aid surgical planning. In normal patients the calibre of the rectum is equal to or greater than the rest of the colon, conversely in HD there is classically a narrow distal segment, funnel-shaped transition zone and dilated ganglionic bowel proximally. In classic segment disease the transition zone is seen at the recto-sigmoid (Fig. 2). The investigation is less sensitive for detecting short segment disease or total colon aganglionosis. 10% do not have a radiological transition zone and 8% are reported to have transition zone proximal to that suggested radiologically [13]. Anorectal manometry is rarely used in the initial diagnosis phase. In some centres it is used to evaluate functional results after definitive surgery; it measures the rectoanal inhibitory reflex, which is classically absent in HD, leading to elevated anorectal pressure [1]. Rectal biopsy is the gold standard for the definitive diagnosis of HD. Indications include failure to pass meconium within the first 48 hours of life, colonic atresia, and recurrent necrotising

<table>
<thead>
<tr>
<th>Neponate</th>
<th>Older child</th>
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<tr>
<td>Failure to pass meconium in first 24-28 hours life</td>
<td>Chronic constipation</td>
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<tr>
<td>Abdominal distension</td>
<td>Chronic abdominal distension</td>
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<td>Bilious vomiting</td>
<td>Vomiting</td>
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<td>Neonatal enterocolitis / sepsis</td>
<td>Failure to thrive</td>
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<td>Colonic atresia</td>
<td>Enterocolitis</td>
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**Table 1.** Clinical presentation in neonates and older children

<table>
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<tr>
<th>Differential diagnosis</th>
<th>Neponate</th>
<th>Older child</th>
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<tr>
<td>Malrotation + / - volvulus</td>
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<td>Chronic constipation</td>
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<td>Meconium ileus</td>
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<td>Anorectal malformation</td>
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<td>Intestinal atresia</td>
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<td>Meconium Plug</td>
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**Table 2.** Differential diagnosis in neonates and older children
enterocolitis; in older children chronic constipation in association with vomiting or abdominal distension [14]. Contraindications include clotting disorders and sepsis. Suction rectal biopsies (RSB) can be performed at the bedside in neonates. In older children full thickness/stripe biopsies are routinely performed under a general anaesthetic, both with antibiotic cover. A 2015 systematic review of 5,068 cases found the mean sensitivity of RSB to be 97% and mean specificity 99%. Complication rates were reported at: overall 0.65%; bleeding requiring blood transfusion 0.53%; perforation 0.06%; pelvic sepsis 0.06% and inadequate sample 10%[15]. The biopsy should be taken from the posterior wall; there is currently no consensus on the number or level of biopsies. Three biopsies are usually taken, commencing at 2.5 cm above the anal verge and 3.5 cm above in the older child, and 1 cm more proximal at each sample. It is vital to document the level at which biopsies are taken.

A rectal washout should not be performed for 24 hours post biopsy. Therefore the biopsy should only be performed when the bowel has been successfully decompressed.

**Histopathology**

Samples are sent fresh to the histopathology laboratory for analysis. Hematoxylin/eosin (H&E) is the most widely used stain and with the absence of ganglion cells a diagnosis can be made. To increase diagnostic accuracy other histochemical and immunohistochemical stains are utilised. A 2016 international survey reported the frequency of staining methods to be H&E 75.9%, acetylcholinesterase (AChE) 73.6%, and calretinin 33.3%[16]. Increased number and hypertrophy of AChE positively stained nerve fibres are seen in patients with HD [10,18]; however, this has been recently questioned [19]. Interpretation of AChE stain requires experience and can be person dependant; therefore there is risk of misinterpretation and false-positive results [20]. Calretinin staining is less widely used; in HD the normal calretinin staining of the intrinsic nerves of the muscularis mucosae and lamina propria is lost, therefore there is less risk of misinterpretation leading to false positive results. [19]

**Initial Management**

The aim of initial management is stabilisation of the patient and decompression of the bowel. The child should be kept nil by mouth, IV access and bloods obtained, nasogastric tube passed and placed on free drainage with ml for ml replacement of losses (0.9% saline and 10 mmol of potassium chloride in 500 ml) and commenced on maintenance fluids. Once decompression has been successfully achieved feeds can be reintroduced. A digital rectal examination or hegar calibration is usually performed prior to commencing washouts. A widebore tube is placed in the distal bowel and volumes of 10-20mls of normal saline, on average 20ml/kg is used to
washout stool and vent flatus, larger volumes can be used if fluid is not retained. If washouts are successful, many centres train the parents to perform washouts to enable discharge and elective planning of the pull-through procedure. If unsuccessful, a stoma is formed to facilitate decompression.

If concern regarding enterocolitis is present patients should be commenced on antibiotics whilst undergoing washouts. Oral metronidazole or vancomycin can be used prophylactically to prevent recurrent episodes [11].

**Definitive management**

The definitive management of HD is surgical. The aim of surgery is to achieve faecal continence through voluntary bowel motions. This requires normal anal sensation, voluntary anal sphincter control, bowel motility [21] and patient compliance in the long-term. Surgical techniques should aim to preserve the first three.

All surgical techniques adopt a pull-through approach, which involve resection of the aganglionic segment of bowel and the TZ and anastomosing the ganglionic bowel to the rectum. The three main pull-through procedures performed are the Swenson, Soave and Duhamel. To date there has been no study showing that one procedure is superior to the others. All can be performed as a one stage or multi stage procedure.

It is important to consider long segment disease as most surgeons would perform the pull-through at a later age in TCA. Biopsies must be performed to identify the TZ and location of the ganglionic bowel suitable for pull-through and anastomosis. They can be performed at the time of stoma formation, prior to or at the time of definitive surgery. The transition zone follows an asymmetric course with some portions of agangliosis extending more proximally. A single-point biopsy may miss the TZ. Therefore it is recommended to sample from all 4 quadrants of the bowel circumference or the entire circumference of proximal resection margin for histopathological analysis [18].

The Swenson procedure was the original pull-through procedure first performed in 1948. It involves dissecting outside the rectal wall. The aganglionic bowel is resected and ganglionic bowel anastomosed above the dentate line. This procedure went out of favour due to the risk of damage to the autonomic nervous plexus and pelvic vessel injury [13]. Recently it has gained in popularity again since it was reported that careful dissection close to the rectal serosa has a low complication rate [22].

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The Swenson procedure involves dissection within the submucosa for most of the dissection, beginning above the dentate line. The remainder of the dissection includes the muscular layer. This leaves an aganglionic muscle cuff through which the proximal ganglionic colon is pulled and anastomosed. As this led to obstructive symptoms the method has been modified and most surgeons use a short cuff or split it.

In the Duhamel pull-through procedure the dissection is performed retro-rectal to avoid pelvic nerve or vessel damage. The ganglionic bowel is anastomosed posterior to the native rectum and a stapling device is used to divide the bowel side-to-side and form a rectal pouch. This leaves a segment of aganglionic bowel behind which contributes to a higher risk of long-term constipation and enterocolitis.

The Swenson and the Soave procedures can both be performed transanally. All three procedures can be performed laparoscopically assisted, to aid dissection and mobilisation of the colon and to facilitate intraoperative biopsies [23].

**Complications/long-term management**

Constipation, faecal incontinence and enterocolitis are the commonest reported long-term complications [24] and can be serious problems to manage. If not managed effectively this can have a significant impact on the child’s physical, emotional and social development.

An evaluation and treatment algorithm can be used for patients who are not progressing well post pull-through to establish the aetiology of their symptoms and guide management. Patients are categorised into patients who soil and patients with enterocolitis, distension or failure to thrive. For patients who soil, the anal canal and sphincter should be assessed. If intact, medical management can be considered, for example loperamide, constipating diet + pectin if the bowel is hypermobile or laxative if the bowel is hypomobile. When the anal canal or sphincter has been compromised enemas, loperamide and constipating diet can be trialled.

Enterocolitis, distension and failure to thrive patients are more likely to present with obstructive symptoms. In these cases an anatomical and pathological cause should be sought. Anatomical anomalies include stricture, after Duhamel procedure megarectal pouch or spur, soave cuff (wide pre sacral space on lateral imaging), dilated retained distal segment and kink or twist to the pulled through bowel. Pathological causes to be considered include aganglionic or TZ pull through [21, 23]. If any of these are present surgical correction is required and after consideration a redo procedure may be warranted.
Table 3. Ano-rectal malformation classifications

<table>
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<tr>
<th>Anorectal Malformations</th>
<th>Classification</th>
<th>Krickenbeck classification</th>
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<tr>
<td><strong>Introduction</strong></td>
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<td>The term Anorectal Malformations (ARM) is used to describe a spectrum of malformations of the anus and rectum, ranging from babies with minor isolated malformations to sick babies with a complex defect and associated anomalies. The anomalies are different in boys and girls. The incidence is approximately 1:5000 life births with a male predominance of about 60%.[25].</td>
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<td><strong>Aetiology</strong></td>
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<td>Anorectal malformations result from an arrest in the process of cloacal separation. The precise causes of this developmental arrest are not clear. It has been suggested that the pathogenesis lies within the homeobox and sonic hedgehog signaling pathways.[26]. Genetic and environmental factors most likely have an influence.[27,28,29]. Several syndromes with a known genetic abnormality include ARM as part of the syndrome, such as Currarino syndrome, Down syndrome or Cat eye syndrome.</td>
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<td><strong>Associated anomalies</strong></td>
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<td>Associated anomalies occur in up to 70% of babies and vary in different regions. Urogenital anomalies account for up to 80%, followed by musculo-skeletal, gastrointestinal tract, cardiovascular, central nervous system and chromosomal anomalies.[30,31,32]. Most common is the VACTERL association (vertebra, anorectal, cardiac, tracheo-esophageal, renal, limb). The “higher” and the more complex the ARM as is the rate of associated anomalies, especially genito-urinary and sacral/spinal.[23].</td>
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<tr>
<td><strong>Classification</strong></td>
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<td>Different classifications have been used to describe the different anomalies. In the past the Wingspread classification divided ARMs into low, intermediate and high lesions. Peña classified according to the anatomical end of the rectal fistula. In 2005 an expert group met in Krickenbeck and agreed on a new International diagnostic classification system, Table III [33,34]. Related anomalies are rectal atresia, which is thought to have a different pathogenesis, resulting from a vascular accident and rectal stenosis. Anorectal malformations should not be confused with a true anterior anus. This is an anus lying more anterior than usual with a normal anal canal within the anal sphincter.</td>
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<td><strong>Types of ARM Female</strong></td>
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<td>Perineal fistula</td>
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<td>The rectum opens partly or completely anterior to the sphincter muscle onto the perineal skin. It is frequently stenotic.</td>
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<td>Vestibular fistula</td>
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<td>The rectum opens within the vestibulum. Three separate orifices are visible at inspection (urethra, vagina, rectum). This is the most common ARM in females.</td>
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<tr>
<td>Cloaca</td>
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<td>The length of the common channel varies. There is one common opening onto the perineum. The length of the common channel varies. The anatomy of cloacas is variable, e.g. the rectum might be inserting high, or there might be a double uterus. About 50% of neonates present with a hydrocolpos.[24].</td>
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<td><strong>Types of ARM – male</strong></td>
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<tr>
<td>Perineal fistula</td>
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| Like in females the rectum opens partly or com
pletely anterior to the sphincter muscle onto the perineal skin. It is frequently stenotic and can be covered (“bucket handle” anomaly or covering membrane).

**Rectourethral fistula**
The rectum ends in a narrow fistula to the urethra. The level of this is variable; the fistula can either be at bulbar or prostatic level.

**Rectovesical fistula**
The rectum ends with a fistula to the bladder neck. In 4-5% of female and male patients no fistula is present. This increases in patients with trisomy 21 and ARM, in whom 95% have no fistula [35].

**Presentation**
Anorectal malformations are most frequently diagnosed at the neonatal check examination or secondary to another malformation such as oesophageal atresia. Babies with perineal fistulas usually pass meconium, unless the fistula is very narrow. If the unusual appearance of the perineum is missed in the neonatal period, children may present late with constipation and abdominal distension. The same is true for girls with vestibular fistula who pass meconium from the introitus.

The most severe form of ARM in girls is a cloaca. Apart from a single opening in the perineum these babies might present with an abdominal mass, usually secondary to hydrocolpos. In boys with a low rectum, meconium might become visible within the first 24 hours of life at the scrotal raphe or penis. Boys with rectourethral or rectovesical fistula develop abdominal distension early and might pass meconium via urethra.

It is important to inspect the perineum and buttocks. Flat perineum and buttocks are a sign of poor muscle and indication of a poor prognosis regarding continence.

**Early Management**
The most important factors when presented with a neonate with ARM are the evaluation for potential life threatening associated anomalies (especially cardiac, oesophageal atresia / trachea-oesophageal fistula) and to decide if a diverting colostomy or primary repair of the lesion should be performed. In the majority of cases the safest approach is the initial diversion of the faecal stream via descending colostomy and corrective surgery performed at a later date.

A primary repair is suitable for most perineal fistulas in female and male neonates. It might not be clear at birth if a baby has a low ending rectum. The decision which operation is performed in these cases is made after 24 hours, when it is clear if meconium has become visible. A cross-table lateral x-ray with the baby in prone position and the pelvis elevated for 15 minutes before the x-ray is taken might help decision making [23].

**Decision making for initial surgery:**

**Males**
- Perineal fistula -> Anoplasty
- All other anomalies -> Colostomy

**Females**
- Perineal fistula -> Anoplasty
- Vestibular fistula -> Colostomy
- All other anomalies -> Colostomy (cloaca +/- draining of hydrocolpos and urinary tract)

Different types of anoplasty are performed for perineal fistulas: YV or cutback anoplasty, minimal posterior sagittal anorectoplasty (PSARP) [32,36]. Some experienced surgeons perform primary PSARPs in vestibular fistulas [36], some keeping the baby on TPN and nil by mouth for up to 7 days.

The colostomy should be placed in the descending colon. This leaves enough bowel distally to later perform the definitive pull-through and has a low incidence of stoma prolapse. Most surgeons prefer a divided stoma, so that stool is completely diverted and does not spill over into the distal colon. This helps to prevent urinary tract infections and allows to use of the mucous fistula to wash meconium completely out and perform a colostogram [37].

**Further Investigations**
All neonates with ARM should be investigated.
for associated anomalies. This includes review by a cardiologist / echocardiography, x-rays of the vertebral column including the sacrum, an ultrasound of kidneys / urinary tract and the spine, looking especially for tethered cord and a pre-sacral mass.

After formation of a colostomy a distal colostogram needs to be performed in all male babies and in female babies without fistula. If contrast is inserted under pressure into the mucous fistula this shows the position of the rectum and fistula. In addition the colostogram allows assessment of the length of distal bowel to determine if this is sufficient to perform the pull-through. In boys it is best performed at the same time as a cystourethrogram to optimise the visualisation of the fistula and assess for associated urinary tract anomalies such as vesico-ureteric reflux (Fig. 3).

In girls with cloaca it is mandatory to perform a detailed cloacogram to evaluate the anatomy before planning definitive surgery. Endoscopy of the common channel needs to be performed to measure the length of the common channel, the urethra proximal (from bladder neck to the common channel) and assessment of the anatomy of Muellerian structures.

Recently the use of MRI for pre-operative assessment of ARM has been advocated in some centres [38].

Urodynamics should be considered pre/ post definitive surgery to assess the operative impact on bladder function.

**Definitive surgical management**

A variety of surgical approaches exist to repair anorectal malformations. Since Peña described the posterior sagittal anorectoplasty (PSARP) in 1982 [33] this has been the most widely performed operation. Some surgeons perform anterior anorectoplasties [39] or a laparoscopically assisted anorectoplasty (LAARP) [40,41]. If a colostomy is performed as initial operation the PSARP is scheduled after all other investigations have been performed and the child is growing well. This is usually the case by 2-3 months of age.

In females the posterior sagittal approach alone is sufficient to repair vestibular fistulas and the majority of cloacas. In the remaining cases a high rectum and/or vagina require an abdominal approach as well. In males high rectourethral (prostatic) and rectovesical fistulas require a combined posterior sagittal and abdominal approach. The abdominal component can be performed laparoscopically or via laparotomy [23].

**Post-operative management**

The urinary catheter that was inserted during PSARP should remain in situ postoperatively to keep the wound dry, thus allowing better wound healing. In males it allows the closed fistula to the urinary tract to heal. In our centre we remove the catheter after 7 – 10 days. Anal calibration / dilatations can be started when the catheter is removed. The size used depends on the size of the neo-anus created and the final size according to the age of the child. Different regimes regarding the frequency of dilatations and the speed of increasing the size exist. We use once daily dilatations, increasing the size as tolerated every 1 – 2 weeks. Once an adequately sized anus is reached and can be dilated with ease the colostomy is closed. It is important to warn the parents that severe perianal excoriations will occur, as the perianal skin has never been exposed to stool before. These are treated with various barrier creams.

**Prognosis**

The prognosis regarding bowel function is dependent on the type of ARM and associated sacral and spinal anomalies. The “higher” and more complex the ARM, the worse the prognosis is for continence, especially in association with sacral and spinal anomalies like tethered cord. An early predictor of continence is the sacral ratio, calculated on a pelvic x-ray [32]. Patients with perineal and vestibular fistula more commonly develop constipation.

Peña reported voluntary bowel movements in 75% of patients, with 50% having occasional soiling. 40% never soil and are completely continent. 25% of patients are incontinent and require bowel management. 43% of patients were constipated [42]. Most studies review smaller numbers of patients and a recent systematic review reported a wide range of long-term problems, including fecal incontinence 16.7% to 76.7% and chronic constipation 22.2% to 86.7%. Other long-term problems included urinary incontinence, ejaculatory dysfunction, and erectile dysfunction [43].

The results reported comparing LAARP to PSARP are similar regarding faecal incontinence and constipation. Posterior urethral diverticula were more common after LAARP [44,45].

**Long-term management**

A very important factor in the treatment of patients with ARM is the long-term management of constipation / soiling, urinary incontinence / neuropathic bladder, gynaecological abnormalities, psycho-social problems and other associated anomalies. Children with lower anomalies (perineal fistula, vestibular fistula) more frequently devel-
op constipation whereas children with higher anomalies have a higher risk of developing faecal incontinence [23]. Children need to be monitored for constipation and treated with laxatives early. If constipation is severe they frequently develop overflow incontinence with regular soiling. This should be treated differently from true faecal incontinence as successful treatment of the constipation stops the soiling. Different laxative regimes, depending on stool consistency and frequency, and dietary advice are often sufficient treatment. In children with true faecal incontinence bowel management with washouts is required. Traditionally this was performed as antegrade continence enema (ACE) after formation of an appendicostomy [ ], but recently successful bowel management has been achieved with Peristeen washout per rectum [47,48].

The majority of children with anorectal malformations achieve urinary continence. If sacral and spinal cord anomalies are present they might develop incontinence or a neuropathic bladder. In girls with cloaca urinary incontinence is more frequent. These children might require intermittent catheterization and should be treated together with a paediatric urologist. In the worst cases these children develop end stage renal disease and the need for renal transplant [49].

17% of girls with vestibular fistula have gynaecological abnormalities such as a vaginal septum, vaginal atresia, or absent uterus [50]. Girls with cloaca need to be investigated for gynaecological anomalies before performing definitive surgery. Most surgeons advocate to deal with these at the same time as definitive surgery is undertaken (e.g. vaginal replacement, division of vaginal septum). All girls who had a PSARP performed should receive advice from a gynaecologist around puberty regarding mode of delivery [51].

Throughout childhood children with congenital colorectal anomalies might develop psychosocial problems [52]. It is therefore useful to offer psychological support to these families as well as liaison with school nurses or other health professionals in the community. It is increasingly recognised that these children frequently require a well organised transition to adult services [53].


32. 2007; 26:2-33.


34. 2004; 33:194-204.


42. 2015; 4(1): 35-47.


47. 2015; 33(12): 2281-95.


49. 2015; 33(12): 2281-95.


52. 2015; 33(12): 2281-95.

53. 2015; 33(12): 2281-95.