INTRODUCTION

Umbilical disorders are frequently encountered by pediatric surgeons. In the newborn, the umbilical cord typically desiccates and separates within three weeks, leaving a dry, “star-like” central abdominal scar that forms the umbilicus. Failure of the umbilical ring to completely close can result in an umbilical hernia – by far the most common umbilical disorder. Discharge or abnormal tissue from the umbilicus is most often due to an umbilical granuloma, but can result from incomplete involution of the urachus or omphalomesenteric duct. Any discharge, mass, or sinus tract is pathological and should be appropriately evaluated and treated. These and other umbilical disorders are discussed in further detail in this review.

ANATOMY AND PATHOLOGY

The umbilical cord is the main portal for entry and exit of blood from the placenta to the fetus during intrauterine life. In addition to the paired umbilical arteries and umbilical vein, the umbilical cord also contains the vitelline or omphalomesenteric duct (which connects the yolk sac to the midgut) and the allantois (the portion connecting the umbilicus to the bladder becomes the urachus). Usually the vitelline duct obliterates by the 5th-9th week of gestation, and the urachus obliterates to become the median umbilical ligament by the 4th-5th month. After birth, the umbilical cord withers and separates, leaving no remnants. Umbilical abnormalities can arise when embryological remnants persist or fail to completely involute (1,2).

The table below compares the embryological components of the umbilical cord with related disorders.

<table>
<thead>
<tr>
<th>Embryological element</th>
<th>Normal remnant</th>
<th>Pathological abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Two arteries</td>
<td>Two arteries</td>
<td>Single umbilical artery</td>
</tr>
<tr>
<td>One vein</td>
<td>Round ligament of liver</td>
<td>Phlebitis</td>
</tr>
<tr>
<td>Allantois</td>
<td>Median umbilical ligament</td>
<td>Patent urachus, urachal cyst or sinus</td>
</tr>
<tr>
<td>Vitelline duct</td>
<td>None</td>
<td>Omphalomesenteric duct remnant, umbilical polyp, Meckel’s diverticulum</td>
</tr>
<tr>
<td>Umbilical ring</td>
<td>Physiologic closure, fascia covering defect</td>
<td>Umbilical hernia, Omphalocele</td>
</tr>
</tbody>
</table>

Like skin anywhere on the body, the umbilicus may also be affected by a variety of dermatological conditions, such as hemangiomas, dermoid cysts, or mechanical irritation. A number of syndromes, such as the Aarskog, Reiger, and Robinow syndromes, are associated with an abnormal umbilical appearance (3). The umbilicus can also be found in an abnormal position or even absent, as in bladder extrophy.
CLASSIFICATION OF UMBILICAL PROBLEMS

Umbilical problems can be classified based on the etiology of the abnormality:

a. infectious: omphalitis, umbilical vein phlebitis
b. acquired: delayed umbilical separation, umbilical granuloma
c. congenital: omphalomesenteric duct remnant, umbilical polyph, patent urachus, umbilical hernia, dermoid cyst, umbilical dysmorphism
d. neoplastic: rhabdomyosarcoma, teratoma.

SELECTED UMBILICAL PATHOLOGY

Omphalitis

Omphalitis is an infection of the cord stump or its surrounding tissues. It presents most commonly in the newborn; the mean age at onset is 5-9 days, earlier in preterm infants. The risk of omphalitis is increased by a number of maternal factors (prolonged rupture of membranes, maternal infection, amniotitis), factors at delivery (non-sterile or home delivery, inappropriate cord care); and neonatal factors (low birth weight, delayed cord separation, leukocyte adhesion deficiency, neonatal alloimmune neutropenia). Developing countries have a significantly higher incidence (as high as 6%) compared to developed countries (0.7%) (2). In developed countries the recommendation for cord care is dry cord care, based on multiple randomized trials performed in high income countries and summarized in a cochrane review (4). However, in developing countries where the risk of omphalitis is higher, evidence from high income countries does not apply.

Proper umbilical cord care is important in decreasing the incidence of omphalitis and neonatal tetanus (which may or may not be associated with omphalitis). Public health interventions have proven effective in decreasing the incidence and death from these infections. In Nepal, the use of chlorhexidine gluconate 4% in aqueous solution decreased the incidence of omphalitis by 75% and its mortality by 24% when compared to dry cord care in a randomized controlled trial involving over 15,000 newborns (5). Soap and water cord care did not significantly decrease either omphalitis or neonatal mortality in a third arm of this same trial. It has been estimated that more than a half million deaths occur yearly in newborn infants from neonatal tetanus (11). A high rate of neonatal tetanus was seen among the Maasai people in Kenya and Tanzania who applied cow dung to the umbilical stumps of their infants. In one simple health program among the Maasai people, the death rate from neonatal tetanus decreased from 82 per 1000 in control groups to 0.75 per 1000 in the intervention group (11). Part of their success was in finding solutions that were culturally applicable and feasible (e.g., if clean water was unavailable, they advocated cleaning the stump with milk), obtaining support from within the community, and maintaining continued health promotion. Other agents that have been used include 70% alcohol, silver sulfadiazine, chlorhexidine, neomycin-bacitracin powder, and salicylic sugar powder (12, 13).

Patients with omphalitis present with erythema, edema, and/or purulent drainage from the umbilical stump. Patients may also have systemic signs of sepsis, including lethargy, irritability, poor feeding, and fever or hypothermia. More extensive disease is seen with necrotizing fasciitis or myonecrosis and may also include a rapidly progressive cellulitis, peau d’orange appearance, violaceous discoloration, bullae, crepitus, and petechiae.

Patients with omphalitis should be admitted and blood and wound cultures should be obtained. Omphalitis is usually polymicrobial and intravenous antibiotics covering gram-positive and gram-negative organisms should be initiated and the area of cellulitis marked and closely followed. Some authors also advocate anaerobic coverage; it certainly should be instituted if there is a concern for necrotizing fasciitis. Newborns with sepsis should also have a lumbar puncture and supportive care instituted.

Patients with necrotizing fasciitis or myonecrosis require emergent and complete surgical debridement of all affected tissue, including preperitoneal tissue, the umbilical vessels, and the urachal remnant. Necrotizing fasciitis or myonecrosis can rapidly progress over a few hours; early and aggressive surgical treatment is critical to survival.

Complications of omphalitis include umbilical phlebitis, portal vein thrombosis (which may lead to portal hypertension), liver abscesses, peritonitis, and necrotizing fasciitis or myonecrosis. The overall mortality of omphalitis is estimated at 7-15 % and is significantly higher (37-87%) if complicated by necrotizing fasciitis or myonecrosis (14).

Delayed umbilical separation

The timing of umbilical cord separation may vary depending on ethnic background, geographic location, and method of cord care. Cord separation usually occurs one week after birth and persistence beyond three weeks is generally considered delayed. However, various umbilical cord antiseptics can prolong the separation time – for example, triple dye may prolong separation of the cord for up to eight weeks. Dry cord care has been found to be effective in developed countries (15), however in developing countries, antiseptic cord care is recommended, and has been found to decrease the incidence and mortality from omphalitis as described above (16).

Aside from agents used in umbilical cord care, other factors that can delay umbilical cord separation include infection, underlying immune disorders (such as leukocyte adhesion deficiency), or an urachal abnormality (17, 18, 19).

On examination, the skin surrounding the umbilical cord remnant should be carefully examined for an urachal remnant or for any evidence of infection, as omphalitis (discussed below) can be rapidly progressive and life-threatening in a neonate.

A complete blood count with a differential may be useful as an initial screen for leukocyte adhesion deficiency. Even in the absence of infection, leukocytosis and neutrophilia may be present in patients with leukocyte adhesion deficiency (10). Rare neutrophil motility defects may require a more sophisticated immunologic workup.
If a patient presents with delayed separation of the cord, it may either be gently removed manually or divided just distal to normal skin with scissors or a scalpel. After removal, the stump site should be cleansed with an antiseptic agent and exposed to air.

**Umbilical granuloma**

This is the most frequent cause of “wet umbilicus,” and presents as moist, raw, reddish-pink tissue arising from the base of the umbilicus after umbilical cord separation. An umbilical granuloma typically measures 0.1 - 1cm in size and may be pedunculated. It is non-tender (lacking innervation). Drainage may be clear or have the appearance of a fibrous exudate. The tissue is friable and may bleed easily. An umbilical granuloma is due to the persistence of capillary and fibroblast cells, markers of an ongoing tissue growth. It may be difficult to distinguish from an umbilical polyp (discussed later), which is usually brighter red, slightly larger, and represents remnant omphalomesenteric duct or urachal tissue.

Management options include repeated cauterization with silver nitrate, ligation, use of alcoholic wipes, or rarely surgical excision. Care must be taken in applying silver nitrate, as contact with normal skin can cause a chemical burn. If the lesion fails to resolve with silver nitrate, the diagnosis should be questioned as umbilical polyps may look similar to umbilical granulomas but do not respond to silver nitrate. If the lesion is excised, histology should be performed to rule out retained omphalomesenteric duct or urachal remnants, which require further work-up.

**Dermoid cyst of the umbilicus**

This is a rare umbilical mass caused by inclusion of skin epithelium below or within the normal skin of the umbilicus. On examination, the umbilicus appears wider, darker in color, and shiny. No inflammation is noted unless the cyst is infected. The diagnosis is made at surgery on finding the characteristic toothpaste-like sebaceous material within the umbilical mass. Surgical excision is curative.

**Omphalomesenteric or vitelline remnant**

During early fetal development, the vitelline or omphalomesenteric duct serves as a conduit from the yolk sac to the midgut. It normally completely involutes by the 9th week of fetal life. However, a portion of or all of the duct may fail to involute and presents as one of the following:

a – an umbilical polyp (see below).

b – a Meckel’s diverticulum, in which only the diverticulum attached to the ileum has failed to involute. This is the most common vitelline remnant; it most often presents as a lower GI bleed caused by ectopic gastric mucosa, but rarely may present as diverticulitis or may function as the lead point for an intussusception.

c – a persistent congenital band, which can act as a fixed point around which an intestinal volvulus may occur.

d – a complete omphalomesenteric duct remnant with a patent conduit connecting the umbilicus to the ileum; this usually presents with pink mucosa protruding from the umbilicus and usually minimal but persistent discharge of intestinal contents or stool.

e – an omphalomesenteric duct cyst, in which the proximal and distal ends have obliterated but a remnant persists in-between; this may present with an infection or obstruction, and is quite rare.

Diagnosis is generally made on physical exam. An ultrasound may show a loop of bowel present under the umbilicus, but is not diagnostic and usually not necessary. A fistulogram may be helpful in clarifying the diagnosis. All omphalomesenteric duct remnants should be surgically resected. A Meckel’s diverticulum should be amputated at its base and the intestine closed.

**Umbilical polyp**

An umbilical polyp is a round, reddish mass at the base of the umbilicus that is comprised of embryologic remnants of the omphalomesenteric duct or, less commonly, of the urachus. It is often brighter red and slightly larger than an umbilical granuloma. Unlike a granuloma, it does not respond to silver nitrate and must therefore be surgically excised and histologically evaluated to confirm the diagnosis. If an umbilical polyp is diagnosed, further work-up for an underlying omphalomesenteric duct or urachal remnant is warranted. One author reported a 30-60% chance of finding an underlying omphalomesenteric duct anomaly if an umbilical polyp was identified.

**Urachal anomalies**

In the fetus the urachus is the embryonal duct connecting the dome of the urinary bladder to the umbilical ring, and is normally obliterated prior to birth, forming the median umbilical ligament. It forms in the peritoneal space between the transversalis fascia and the peritoneum. Non-closure of the entire tract leads to a patent urachus, while closure on the bladder side creates an umbilical sinus. Closure of both ends but patency of the tract between may trap fluid in an urachal cyst, which is the most common urachal anomaly. A bladder diverticulum results when the distal tract involutes, and is the rarest urachal anomaly.

Both a patent urachus and a urachal sinus may present with clear drainage from the umbilicus, and careful examination demonstrates a sinus at the base of the umbilicus. A patent urachus drains urine and may predispose to cystitis or recurrent urinary tract infections. A urachal cyst most commonly presents once it has become infected – an affected patient will present with infraumbilical swelling, abdominal pain and erythema. The symptoms may mimic appendicitis. Patients with delayed separation of the umbilical cord may have a urachal anomaly.

Ultrasoundography is often useful in diagnosing an urachal cyst, and will show a cystic hypoechogenic lesion in the pre-peritoneal space. The presence of a longitudinal double line from the bladder dome to the umbilicus is indicative of a urachal remnant. A sinogram may be used to identify the presence of a patent urachus or an urachal sinus. For a patent urachus, a VCUG should be obtained to exclude the presence of posterior urethral valves (back-up pressure from the distal obstruction may be keeping the urachus patent).

Treatment involves complete resection of any part of the tract that has failed to completely obliterate. It is important to remove a cuff of bladder when excising the urachus to prevent the risk of developing an urachal adenocarcinoma later in adulthood.

**Umbilical hernia**

http://www.ptolemy.ca/members/current/Abdomen/index.html
An umbilical hernia is a full-thickness protrusion of the umbilicus with an associated fascial defect, and may contain peritoneal fluid, pre-peritoneal fat, intestine or omentum. Umbilical hernias are more frequent in premature, low birthweight, and black infants. They also occur more often in children with ventriculoperitoneal shunts, ascites, obesity, and certain syndromes including Beckwith-Wiedeman, Down’s, and Marfan’s syndrome (1). An umbilical hernia results when the umbilical ring fails to close.

Umbilical hernias are common in Africa. In one study from Nigeria, umbilical hernias were found in 91% of under-six year olds; 64% of 6-9 year olds, and 46% of 10-15 year old Nigerian children (22). Meier found umbilical hernias with a >1cm fascial defect in 23% of Nigerian children less than 18 years old (23). Surprisingly, when 6-9 year old Nigerian children of high socio-economic class were evaluated for an umbilical hernia, only 1.3% of 7,968 children had an umbilical hernia (24). It is possible that nutrition may be a factor, as Jelliffe found a higher incidence of umbilical hernias in malnourished vs. well-nourished adults (27% vs. 14%) (15).

In children, umbilical hernias often close spontaneously. Small defects (<1cm) are much more likely to close than large defects (>2cm). Meier et al. reported that umbilical hernias continue to close until the age 14 in African children (16). The skin overlying an umbilical hernia may continue to stretch and result in a proboscoid umbilical hernia. In Africa, most parents are very accepting of its appearance, in contrast to developed countries. Once the umbilical defect has spontaneously closed, the nipple-like umbilical skin may continue to flatten even during adolescence.

Complications including incarceration, strangulation, and rupture of umbilical hernias may occur. In developed countries, the incidence of incarceration or strangulation is rare – one paper reported an incidence of one in 1,500 umbilical hernias (25). Rupture of umbilical hernias with evisceration is even more rare, but has been reported in infants < 6 months old (26, 27).

While the incidence of incarceration and strangulation in children with umbilical hernias in Africa is not known, it appears to be higher than in the West (although this may in part reflect the significantly higher prevalence of umbilical hernias in black children). For instance, at A. Le Dantec Hospital in Senegal, over a 5-year period 41 children had emergency operations for incarcerated or strangulated umbilical hernia. At Jos University Teaching Hospital in Nigeria, over an 8-year period 23 children underwent surgery for acute or recurrent incarceration (26, 23).

In contrast, Okada et al. reviewed the literature from 1957-1999 and found a total of only 38 cases reported in children worldwide. In King’s College Hospital in London, only three incarcerated umbilical hernias were treated in children over a 20 year period (and all three of these occurred in black children). The fact that most umbilical hernias in the West are repaired by 4-5 years of age does not account for the apparent difference in the frequency of incarceration between the West and Africa. In both Senegal and Nigeria, most of the incarcerations reported occurred in patients less than five years old: in Senegal the average age at incarceration was 14 months (range 8 months-10 years), and in Nigeria the median was 4 years old (range 3 weeks-12 years). While most incarcerated hernias do not have an inciting factor, bezoars, digested vegetable matter, parasitic worms, or ascites have been implicated (21, 22, 23).

Umbilical hernias can incarcerate regardless of the size of the fascial defect. Okada et al. reported that a majority (52%) of the patients in their review of incarcerated hernias had medium sized (0.5-1.5 cm) fascial defects, while 24% occurred in small defects (<0.5 cm) and 24% in large defects (>1.5 cm) (24). In Nigeria, Chirdan found that of the incarcerated hernias in which a measurement was documented, all had defects greater than 1.5 cm in diameter (not all, however, were measured) (22).

Factors that lead parents to seek medical care for their child in Africa include the age of the child, size of the defect, height of protruding umbilicus, and pain. On examination, a child with an umbilical hernia usually presents with a protrusion of the umbilicus with contents that are easily reducible. After reduction, the size of the fascial ring can be palpated and can range in size from a few millimeters to more than 4 cm in diameter. No other investigations are required for diagnosis.

An umbilical hernia repair is one of the most frequent procedures performed by pediatric surgeons in developed countries (1). In Africa, however, umbilical hernia repairs are more infrequent as they are usually repaired only if symptomatic or complicated. Generally accepted indications for management in Africa compared to the West are highlighted below.

Since there is a high rate of spontaneous closure and the appearance of a proboscoid umbilical hernia is well-tolerated by parents in Africa, conservative management of asymptomatic, easily reducible umbilical hernias is recommended. In the West, conservative treatment is generally recommended for small asymptomatic umbilical hernias (<2 cm fascial defect) in children less than 4-5 years old.

In Africa, surgical repair is reserved for symptomatic umbilical hernias. Rarely, parents may request to have a proboscoid hernia repaired. (For further discussion, see the section on “ethical issues”.) In the West, surgical repair is generally recommended for hernias with large fascial defects (>1.5-2 cm), hernias that have failed to spontaneously close by 4 to 5 years of age, and umbilical hernias with significant proboscoid components.

A classic Mayo “vest-over-pants” procedure or simple approximation with long-lasting absorbable suture are both acceptable for a conventional umbilical hernia repair. For complicated umbilical hernia repairs, the use of mesh may be considered in the closure of a very large uninfected umbilical hernia to prevent excess tension on the fascia. The use of mesh also prevents the development of an abdominal compartment syndrome which could result with significant fascial tension. Surgical complications are rare after umbilical hernia repairs. The outcome is excellent and the mortality approaches zero for elective repairs. Rare post-operative eviscerations can be prevented by meticulous surgical technique.

Other umbilical problems

Absent umbilicus: Malposition or absence of the umbilicus is encountered frequently in patients with bladder extrophy. When the umbilicus is absent, an omphaloplasty may be performed, as many ethnic groups are culturally sensitive to the absence of the navel. Research has been performed to help the reconstructive surgeon locate the umbilicus in an aesthetically pleasing location.

Stoma at umbilicus: Some pediatric surgeons in the West have advocated placing intestinal or urinary stomas in the umbilicus primarily for aesthetic considerations. Experience with this in Africa has been limited, and no papers adopting its use have been published in Africa to date.

Ethical Issues

As previously discussed, most African surgeons do not use the same indications for umbilical hernia repair as in developed countries. Instead, they...
recommend repairing only umbilical hernias that are symptomatic in children. However, the incidence of incarceration or strangulation seems to be higher in Africa than in developed countries. Because of this, some African surgeons have recommended repairing all umbilical hernias in children (21). Others, however, continue to recommend conservative treatment in spite of the risk of incarceration (16, 22). Part of the rationale given was the wide prevalence of umbilical hernias. Even using selective criteria, Meier et al. estimated that if umbilical hernias >1.5 cm were repaired in young children in Africa, about 6-8% of children less than four years old would require repair. The exact criteria for elective repair that these estimates were based on was for females greater than 2 years old and males greater than 4 years old with a fascial defect greater than equal to 1.5 cm in diameter; he estimated that 6% of 2 year-old females and 8% of 4 year-old males would need repair. (16) The volume of cases would likely outstrip available surgical resources.

If hernias with large (>1.5 cm fascial defects) are indeed the most likely to incarcerate in Africa, as reported by Chirdan et al. (22), it could be argued that they should be repaired: they are the most likely to incarcerate and the least likely to spontaneously close. Consideration should also be given to closing umbilical hernias in patients who live more than one to two hours away from surgical resources. More research is necessary to determine the actual incidence of incarceration or strangulation, and to clearly define which umbilical hernias are at greatest risk.

Perhaps it is time to re-examine how current recommendations for umbilical hernia repair in Africa were developed or became generally accepted. Are the current recommendations truly the best for the patient, preventing many children with umbilical hernias from unnecessarily undergoing the risk of a surgical procedure and anesthesia? Or did the current recommendations arise out of necessity, due to the wide prevalence of umbilical hernias, in an effort to strategically utilize surgical resources and time? If the latter is true, and umbilical hernias should be repaired using the same criteria as in developed countries, there may be other creative solutions. For instance, just as people without full surgical training have been specifically trained to perform simple, straightforward surgical procedures like umbilical hernia repairs with an appropriate level of supervision.

Key Summary

- Appropriate umbilical cord care is important in preventing omphalitis, which can be a life-threatening infection.
- Randomized controlled trial evidence favours chlorhexidine gluconate 4% for cord care in low income countries.
- The presence of a remnant of the urachus or omphalomesenteric duct should be considered if there is an umbilical sinus, persistent drainage, or remnant tissue.
- Ultrasonography can be useful in investigating umbilical disorders when the diagnosis is uncertain.
- Any irreducible umbilical mass or persistent umbilical lesion needs surgical exploration and resection.
- Umbilical hernias are common in African children, and the incidence of incarceration appears to be higher than in the West.
- In Africa, profusoid umbilical hernias are common, well-accepted, and treated conservatively.
- Incarceration or strangulation of umbilical hernias is uncommon but has been reported more often in Africa than in developed countries.
- Complications remain the general indication for umbilical hernia repair in Africa.
- An omphaloplasty may be indicated for cultural reasons.
- The prognosis is excellent and complications are rare after umbilical surgery.

References


1Paediatric Surgery fellow (COSECSA); 2,3Consultant Paediatric Surgeon; BethanyKids at Kijabe Hospital, Kijabe, Kenya

http://www.ptolemy.ca/members/current/Abdomen/index.html

2010/07/30

Save the review in PDF

Click here to join the Surgery in Africa Discussion Group

(Back to Top)