



# Current management of umbilical abnormalities and related anomalies

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## INDEX WORDS

Umbilical hernia;  
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Prenatally, the umbilicus is of paramount importance, providing the gateway between the mother and the fetus. As the fetus becomes increasingly autonomous at the end of the second month of fetal life, the connections (vitelline, urachal) diminish in significance and involute. Disturbances in this process can result in a wide variety of abnormalities, ranging from relatively minor defects identified at birth (umbilical granulation tissue) to life-threatening complications quiescent until late adulthood (urachal carcinoma). This section will review the 'state of the art' in evaluation and management of these umbilical and related abnormalities.

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Most umbilical abnormalities in the newborn present as failure of the umbilical cord to separate, infection, mass, or drainage. The umbilical cord remnant usually separates in the first month of life, and persistence after the end of the second month of life is considered abnormal.<sup>1</sup> Delayed separation can sometimes be due to underlying medical abnormalities, specifically immune disorders, but these are rare.<sup>2</sup> Prenatal identification of umbilical cysts may indicate a significant risk of chromosomal abnormalities, as high as 18/23 cases in one report.<sup>3</sup>

Excess bleeding from the cord can be a sign of an underlying hematologic disease (eg, Factor X deficiency, an autosomal recessive disease equally affecting males and females, but with an incidence of only 1 in 500,000). A single umbilical artery (2 vessel cord) is found in about 1% of singleton (versus 5% in multiple) pregnancies. Most often, this is an insignificant and incidental finding. However, as many as 25% of infants with a single umbilical artery have underlying abnormalities. Cardiac and renal

ultrasound and chromosomal studies, in addition to a thorough history and physical, are usually obtained.<sup>4</sup>

## Mass

The most common mass at the umbilicus is granulation tissue, followed by umbilical polyps (small bright red remnants of intestinal or gastric mucosa). These two may be impossible to differentiate clinically. Granulation tissue will usually respond to silver nitrate therapy. If there is no response after two or three attempts at silver nitrate therapy, surgical excision may be necessary. Pedunculated lesions with a narrow stalk can be managed with ligation of their base with absorbable suture. Older children sometimes present with a firm, nontender, whitish-pink polypoid mass at the umbilicus that proves to be a cicatrix on pathologic examination. The presence of granulation tissue is not a marker for underlying abnormalities, and no further workup is indicated. However, a true polyp may indicate the presence of underlying anomalies. In one report, there was roughly a 30% to 60% chance of another OMD anomaly if an umbilical polyp was noted.<sup>5</sup> Dermoid cysts and other neoplastic lesions may rarely present as an umbilical mass, usually in older patients. Teratomas, rhabdomyosarcomas,

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malignant fibrous histiosarcoma, and a wide variety of rare tumors have been reported in the umbilicus. A rare abnormality is seen in the newborn noted to have a small umbilical remnant in conjunction with a scaphoid abdomen and a bowel obstruction. The child is then found to have had in utero midgut volvulus, and the "remnant" is the residual atrophic intestine. These children rarely survive.

## Infection

Serious umbilical infections in the newborn have become rare in the developed world but still are a common cause of morbidity and mortality in less developed countries. Umbilical infections in the infant are often related to hygiene issues. The incidence is increased in home births. Severe infections such as omphalitis are currently extremely uncommon, probably as a result of standardized application of topical antibiotics to the newborn umbilical cord and observation of routine hygiene principles. Although infrequent, omphalitis can rapidly progress to severe cellulitis or necrotizing fasciitis, and therefore should be treated promptly and aggressively. Portal vein thrombosis is a recognized sequela. These are life-threatening infections with a high mortality rate.<sup>6,7</sup> Broad spectrum antibiotic treatment is provided, and surgical debridement may be necessary. One 20-year review of 140 infants with omphalitis noted that 8/140 developed polymicrobial necrotizing fasciitis.<sup>8</sup>

## Drainage

Most infants with umbilical granulation tissue/polyps will have a small amount of drainage (stains on clothing), but significant drainage, particularly if it resembles intestinal contents or urine, should suggest the possibility of urachal or vitelline remnants. Ultrasonography should be obtained, and a fistulogram/sinogram may occasionally be useful in this setting. Persistent umbilical drainage can signify an underlying abnormality. In one study, 11/18 patients had a patent vitelline remnant, and 1/18 had a patent urachus.<sup>9</sup> An unusual cause of umbilical discharge in older children is pilonidal disease. Most of these patients are young men, who are successfully treated with excision of the umbilicus.<sup>10</sup>

## Umbilical hernia

### Embryology

The fetal vitelline vein is obliterated and becomes a fibrous cord known as the round ligament of the liver. Orda and Nathan found that this round ligament attaches to the inferior portion of the umbilical ring in about three-fourths of individuals, thus providing an additional layer preventing

umbilical hernia (in addition to the transversalis fascia and peritoneum).<sup>11</sup> However, in approximately 25%, the attachment of the round ligament is to the superior border of the umbilical ring, leaving an area of potential weakness. Furthermore, variation can occur in the fascia umbilicus, a thickening of the transversalis fascia at the umbilicus. It may cover only part of the umbilical ring, or be entirely absent. Richet's fascia is a continuation of the transversalis fascia, a fold of extraperitoneal fascia enveloping the obliterated umbilical vein.

Umbilical hernia is markedly increased in premature infants and may be seen in as many as 75% of infants under 1500 g.<sup>12</sup> The rate of spontaneous resolution is high. It is estimated that as many as 10% to 20% of all infants are born with an umbilical hernia; the incidence is increased in infants of African descent, and is increased in association with certain disease states (Beckwith-Wiedemann syndrome, Down's syndrome). There are no significant gender differences.

## Complications

Rupture of an umbilical hernia can occur, but is extremely rare. However, there are several reports from underdeveloped countries noting a relatively high frequency of incarceration and other complications.<sup>13,14</sup> Why a difference based on socioeconomic, geographic, or perhaps genetic factors should exist is unclear. The incidence of incarceration or strangulation from an umbilical hernia in this country is exceptionally rare. We have not had an emergency operation for umbilical hernia in the past 15 years at our institution.

## Treatment

The factors which primarily determine the chance of closure include age and size. In one often-cited study, there was an approximately 80% chance of closure if the defect was under 1 cm in cross-sectional diameter.<sup>15</sup> Another longitudinal report (6 years) found that 96% of defects <0.5 cm closed spontaneously, but no defect >1.5 cm closed in that interval.<sup>16</sup> In 1 prospective study from Nigeria, spontaneous closure of umbilical hernias was still possible up to age 14 years.<sup>17</sup> Our practice is to observe asymptomatic hernias until about 5 years of age. Symptomatic or extremely large defects are treated more promptly. A frequent issue is the asymptomatic infant with a relatively small umbilical hernia undergoing general anesthesia for an unrelated problem (eg, inguinal hernia). We discuss the options with the family, but would usually recommend continued observation of the hernia in children under 3 to 4 years of age with defects smaller than 1 cm. Laparoscopic hernia repairs have been reported. Albanese and coworkers described 41 umbilical and 13 epigastric hernias repaired laparoscopically with no perioperative complications and no recurrences.<sup>18</sup> Pressure dressings are routinely left on after umbilical hernia repair.

One recent randomized study suggests that this may be unnecessary. Ninety-six patients were prospectively randomized to open versus pressure dressing after umbilical hernia repair; there was no significant difference in terms of wound infection, hematoma or seroma formation, and recurrence rate.<sup>19</sup>

## Umblicoplasty

A wide variety of umblicoplasty techniques are available. Even with relatively large amounts of excess skin, most defects can be managed with simple closure and retention of the redundant skin, which is simply tacked down to the fascia. Over time, the skin contracts, leaving a good cosmetic result. Huge defects may require either circular excision of the excess skin near the base (which is then closed in a purse-string technique), or other types of plastic closure.<sup>20,21</sup> Our preference is to retain the skin in most repairs; simple excision and absorbable purse-string suture closure has given very satisfactory cosmetic results in those few children with very large, proboscis-like protuberances.

## Vitelline Abnormalities

### Introduction

What's new in regard to Meckel's diverticulum? A survey of the literature reveals a fairly constant number of publications on the topic; from 2000 until 2005, the number of publications per year ranged from a low of 30 to a high of 42. Nevertheless, improved imaging techniques and the advent of minimally invasive surgery have changed the diagnosis and management of patients with vitelline abnormalities.

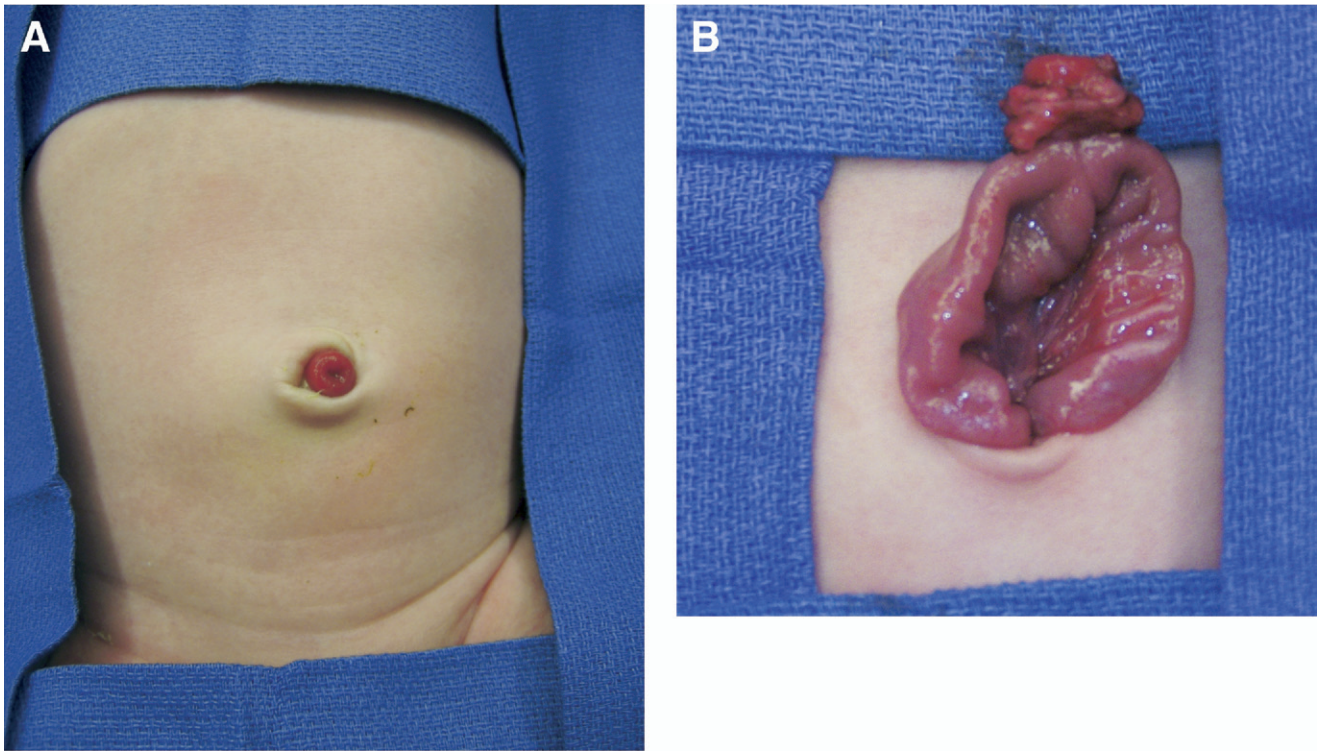
### General

Omphalomesenteric duct (OMD) or vitelline duct anomalies are a group of defects resulting from failure of involution of the OMD, a process usually complete by the 8th to 9th week of fetal life. Underlying genetic causes have not yet been identified. One murine study implicated absence of inhibitory mesodermal interactions during development as a potential cause of Meckel's diverticulum (MD).<sup>22</sup> The sex distribution is equal, but males are more likely to become symptomatic. Although a number of other OMD anomalies can occur, MD is by far the most common. A patent OMD is one of the least common variants (about 5%), and is usually readily identifiable in the newborn period (Figures 1A and B). A small communication sinus tract may require ultrasonography or occasionally a sinogram/fistulogram for diagnosis. Surgical excision is curative. A fibrous OMD tract from the small bowel (usually a MD) to the umbilicus can result in volvulus or internal hernia, or remain asymp-

tomatic. OMD cysts are rare, much less common than their urachal counterparts. They may present with infection or bowel obstruction. Again, resection is curative. The remainder of this section will focus on the most common type of OMD anomaly: Meckel's diverticulum.

### Diagnosis

Most MD are sporadic, but their presence is reportedly increased in children with Hirschsprung's disease, Down syndrome, esophageal atresia, duodenal atresia, malrotation, and congenital cardiac abnormalities. Symptoms vary with age. The most common clinical course for an individual with MD is to remain asymptomatic over one's lifetime. The estimated lifetime risk of symptoms from an MD probably lies in the range of 4% to 6%.<sup>23</sup> The most frequent presentations are obstruction, bleeding, and slightly less commonly, inflammation. Lower GI bleeding from a MD is usually described as brick red or maroon, is painless, and occurs only intermittently. Blood loss may be enough to require transfusion. A Meckel's scan (technetium-99m pertechnetate) is specific for gastric mucosal cells; the accuracy is greater than 90% in children (but lower in adults), and is improved by administration of pentagastrin and H<sub>2</sub> (histamine) blockers. The risk of radiation exposure, particularly in children, is a topic of resurgent interest. It is estimated that a Meckel's scan provides an overall exposure of approximately 0.031 rads, less than from an upper gastrointestinal and small-bowel follow-through study.<sup>24</sup> False-positive results can be due to duodenal ulcer, small-intestinal obstruction, intestinal duplication, ureteric obstruction, aneurysm, and angiomas of the small intestine. False-negative results can occur as a result of necrosis of the heterotopic mucosa, insufficient amount of gastric mucosa within the diverticulum is insufficient, or if intraluminal scintigraphic activity is diluted as a result of brisk hemorrhage or bowel hypersecretion. A child with persistent suspicious bleeding and a negative workup (including a Meckel's scan) may present a diagnostic dilemma. In an otherwise healthy child, consideration should be given to diagnostic laparoscopy as the next step. Many authors advocate this approach.<sup>25,26</sup> In one review of 165 children undergoing Meckel's scan (of whom 70 had proven MD), the false-negative rate of the scan was significant (even if the study was repeated). The authors argued that laparoscopy or celiotomy may be a useful diagnostic tool with a negative scan and a high index of suspicion.<sup>27</sup> Tagged red cell scans, contrast upper or lower GI studies, or even angiography have been recommended, but their accuracy is limited. A solitary, dilated loop of bowel on plain film or CT scan may offer a subtle clue to the diagnosis (Figure 2). Although a logical consideration, helicobacter pylori is not commonly identified in heterotopic gastric mucosa in cases of bleeding MD, even when carefully sought for.<sup>28</sup> Obstruction is another common presentation, with multiple possible mechanisms: intussusception with MD as a lead point, internal hernia beneath a point of attachment to the abdominal wall or vitelline artery,



**Figure 1** (A) This illustrates a patent omphalomesenteric duct. (B) The usual appearance is that of an umbilical stoma. (Color version of figure is available online.)

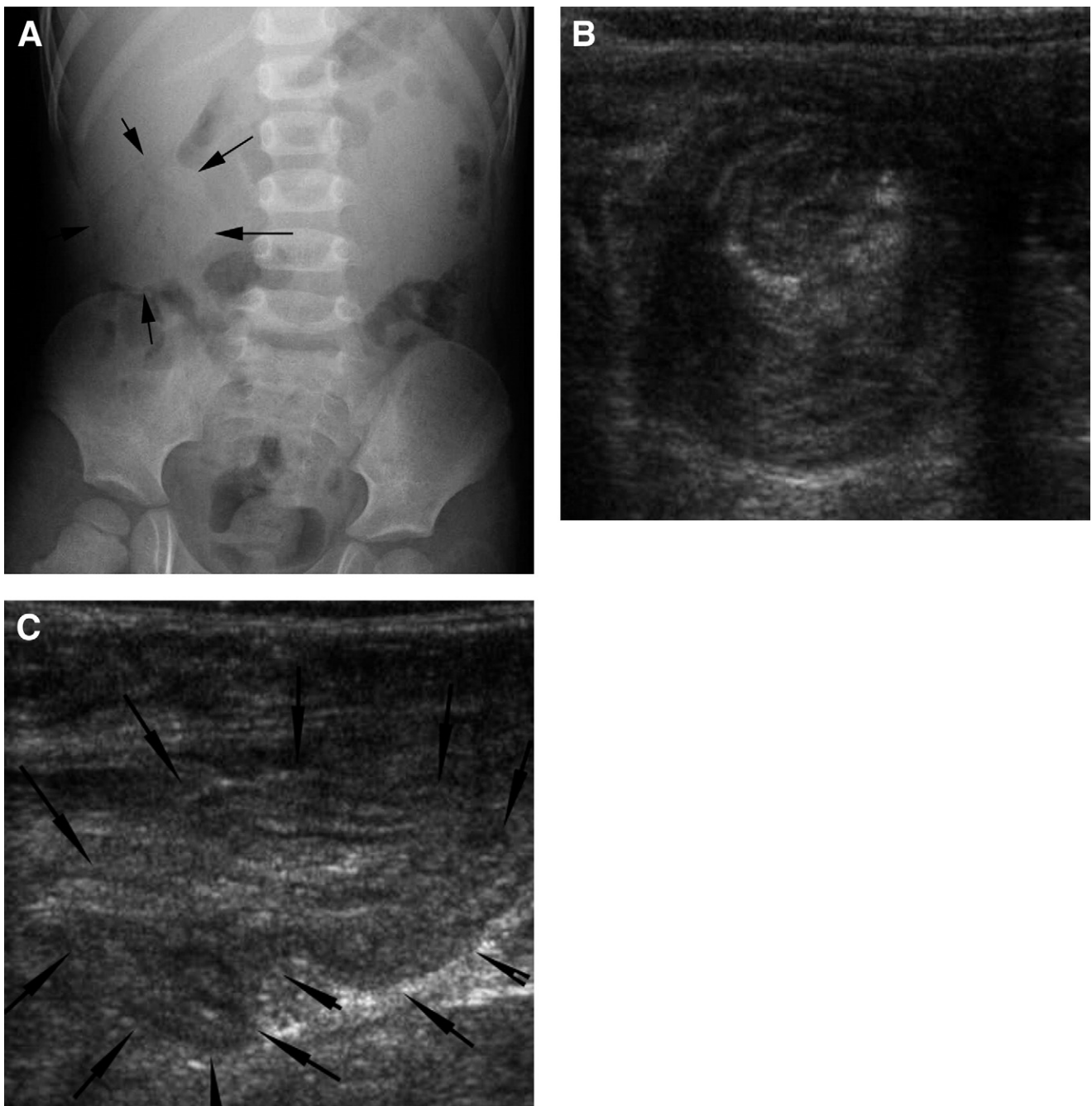
volvulus around an attached band, prolapse through a patent OMD, or secondary to an inflammatory process. MD should be considered when a previously unoperated child presents with a de novo bowel obstruction, or in a child >2 years of age with an intussusception. Most intussusceptions due to

MD are not reducible. Pathologic lead points in intussusception are often due to MD. Fluoroscopy during reduction will miss most pathologic lead points. In one series, nearly two-thirds of such lead points were identified ultrasonographically (Figures 3A–C).<sup>29</sup> However, children with intussusception only very infrequently have an underlying MD. This is true for adults as well; probably 5% to 10% of patients with intussusception outside the “typical” age range, at most, will have MD.<sup>30</sup>



**Figure 2** CT scan of solitary region of dilated bowel with an air fluid level (arrow), suspicious for Meckel’s diverticulum (Courtesy of Dr. Lisa H. Lowe, Children’s Mercy Hospital).

Infection/inflammation in MD is the least common of these three main modes of presentation. Most children with an inflamed MD have heterotopic mucosa; the incidence is not 100%, but necrosis and mucosal obliteration, and degree of diligence in the search for it, may obscure its pathologic identification. Inflammation due to MD is usually diagnosed by CT scan, since the tentative diagnosis is usually appendicitis.<sup>31</sup> Ultrasound is successful in identifying the diverticulum in about half of children with symptomatic (but nonbleeding) MD.<sup>32</sup> Children who are found to have a normal appendix at operation for presumed appendicitis should have their small bowel carefully examined for a MD. Although the “rule of 2” (2% of the population, 2 inches long, 2 types of heterotopic mucosa, symptoms before age 2 years, within 2 feet of the ileocecal valve) is well known, the MD may be located more proximally. In one study, more than one-fourth of MD were located between 91 and 161 cm from the ileocecal valve (from 3 to 5.5 feet).<sup>33,34</sup> Obviously, this may differ with age and total length of small bowel. Other presentations can occur



**Figure 3** (A) A plain radiograph of an intussusception due to a Meckel's diverticulum. There is a paucity of bowel gas in the right upper quadrant, with a plain film "target sign" delineated by the arrows. (B, C) A transverse ("target sign") and longitudinal ("pseudo kidney sign") ultrasound picture in the same patient (Courtesy of Dr. Lisa H. Lowe, Children's Mercy Hospital).

rarely: foreign bodies in MD, parasitic infections, neoplasms, etc. The incidence of tumors within a Meckel's diverticulum is 0.5% to 3.2%, almost always identified in adulthood.<sup>35</sup>

### Treatment

Symptomatic children may require preoperative resuscitation; antibiotics, crystalloid or blood products administration, and bowel decompression may be necessary. Surgical

excision is the treatment of choice for MD. Laparoscopy is increasingly used in both the diagnosis and treatment of MD. The diverticulum can be exteriorized, allowing either diverticulectomy or segmental resection and reanastomosis. Diverticulectomy alone can potentially result in either retained heterotopic mucosa or, in the case of bleeding, a retained ulcer on the mesenteric aspect of the bowel. Seventy years ago, the site of the ulceration in bleeding MD was mapped in 45 patients; most were ulcerated in the neck of the diverticulum.<sup>36</sup> One recent report reviewed 77 patients

with a MD: short diverticula (<2.0 cm) had a wide distribution of heterotopic mucosal sites, with 12 (60%) involving the whole diverticulum including the base, and 8 (40%) involving the tip and body only.<sup>37</sup> It is probably prudent in laparoscopic Meckel's diverticulectomy (particularly in nonincidental scenarios) to deliver the diverticulum extracorporeally, resect, and reanastomose the involved segment, and then return it to the peritoneal cavity.

### Incidental MD

Surgeons should expect to find a Meckel's diverticulum during an appendectomy in about 2% of patients. Ueberueck reviewed 7927 appendectomies over 36 years; 2.9% had a MD.<sup>38</sup> Heterotopic mucosa was found in about 12%. The authors recommended: "Meckel's diverticulum with obvious pathology should always be removed. In cases of gangrenous or perforated appendicitis, an incidentally discovered MD should be left in place, whereas in an only mildly inflamed appendix it should be removed." Another well-known population-based study from Olmstead county supported incidental diverticulectomy.<sup>23</sup> However, other studies caution against incidental diverticulectomy, citing a small but significant risk of anastomotic leak and bowel obstruction<sup>39</sup>: incidental diverticulectomy in 93 adult patients resulted in 2% morbidity but no mortality.<sup>40</sup> Using conditional probability analysis and data from the literature, the authors concluded: "the conditional probabilities of producing surgical morbidity or mortality in the adult population at risk by only resecting symptomatic diverticula are 0.2% and 0.04%, respectively. The comparable risks for resecting all incidentally discovered diverticula are 4.6% and 0.2%. Incidental diverticulectomy in adults should be abandoned." Several authors have suggested that clinical parameters be used to help decide whether or not to perform incidental diverticulectomy (age of patient, palpable or size characteristics of the diverticulum, reason for primary operation). One review of 68 MD compared symptomatic and asymptomatic patients, finding that symptomatic patients had significantly longer diverticuli (>2 cm) and were more likely to have a narrower base than patients who had an incidental diverticulectomy.<sup>41</sup> The palpable characteristics of the diverticulum may be unreliable; in 1 study of 77 adult and pediatric patients, when the MD was described as thickened, heterotopic mucosa was identified slightly less than half the time. Another paper reviewed anatomic distribution of heterotopic mucosa and also found that shorter diverticula were more likely to have involvement at the base, augmenting the suggestion that a segmental resection be performed for short diverticula.<sup>42</sup> Extraneous factors may play a role in the decision-making process: medicolegal concerns if a MD is left in situ but later develops complications/malignancy may be a motive force. The results of several excellent epidemiologic reviews notwithstanding, the decision regarding the management of the incidental Meckel's diverticulum often appears to fall under the

umbrella of "*De Gustibus non est Disputandum*": in matters of taste there can be no dispute.

### Advances

Another new diagnostic tool is wireless capsule endoscopy. This has been used in children as young as 2.5 years to identify MD, and in a wide range of age groups to diagnose other obscure causes of upper and lower GI bleeding.<sup>43,44</sup>

Push-pull endoscopy, first introduced by Yamamoto in 2001, allows endoscopic examination of the entire small bowel from either oral or oral-anal approaches. This has been used in adults to identify Meckel's diverticulum.<sup>45</sup> With this technique, the scope and an overtube are inserted through the mouth and advanced into the small bowel. The scope is advanced a small distance in front of the overtube, and the balloon at the end is inflated. Friction allows the small bowel to be accorded back to the overtube. The overtube balloon is then inflated, and the endoscope balloon is deflated. The process is repeated, allowing the entire small bowel to be visualized.

## Urachal Abnormalities

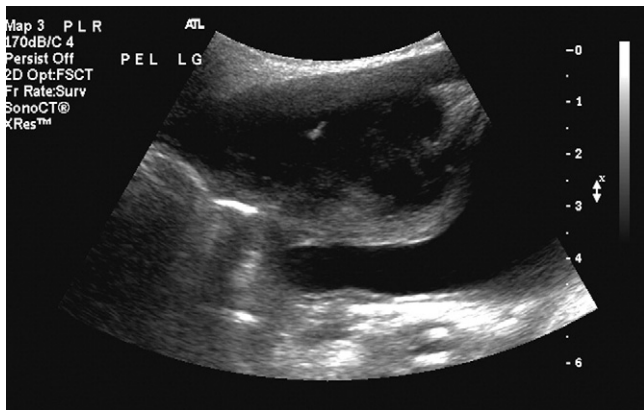
### Anatomy

The urachus connects the bladder to the allantois. The non-patent urachus becomes the median umbilical ligament, usually by about the 4th or 5th month of gestation. Urachal abnormalities can present in many forms. Urachal sinuses can be an incomplete tract, or a complete one (patent urachus). The most common abnormality, urachal cysts, can occur anywhere between the bladder and umbilicus. However, most occur in the distal third of the urachus. Vesicourachal diverticuli are rare, consisting of outpouchings of the bladder at the insertion of the urachus.<sup>46</sup> Most are asymptomatic and incidentally discovered. However, they can be a nidus for infection and stones, particularly if they retain urine.

The underlying etiology at a genetic or molecular signaling level remains unknown. Bladder outlet obstruction is not present postnatally in most urachal abnormalities. Coexistence of a vitelline and urachal remnant is distinctly uncommon, although reported.<sup>47</sup> Heterotopic mucosa, common in vitelline remnants, is unusual in urachal remnants.

### Diagnosis

In one review of 56 children with urachal abnormalities, slightly more than half (36/56) were identified incidentally.<sup>48</sup> A recent review of 45 children with urachal abnormalities (mean age 4 years, and equal gender distribution) found the most common symptoms to be umbilical drainage (42%), mass or cyst (33%), and pain (22%).<sup>49</sup> Urinary symptoms



**Figure 4** This demonstrates a sagittal ultrasound of a urachal cyst (superiorly) with the normal urinary bladder inferiorly and to the right.

were infrequent (<5%). The incidence of associated GU anomalies is low (only 1 of 26 patients in a recent study).<sup>50,51</sup>

Any child suspected of urachal abnormality should undergo ultrasound evaluation as an initial screening test.<sup>52</sup> Drainage from the umbilicus should prompt a contrast sinogram/fistulogram, as well as an ultrasound to identify renal or other GU anomalies (Figure 4). A VCUg is rarely informative in patients with suspected urachal anomalies, with the possible exception of infants with a patent urachus in whom posterior urethral valves are a consideration.<sup>49,51,53</sup>

In infancy, many urachal cysts are identified when an ultrasound is requested to evaluate a baby with umbilical granulation issue. Some patients undergo ultrasound for another indication, and a cyst is a purely incidental finding. Dependent on the clinical history and ultrasound findings (primarily the size of the cyst and the presence or absence of any symptoms), these can be watched or excised. One study addressed this issue; 182 children without any urachal-related symptoms underwent ultrasonography, and “urachal remnants” were found in 99%. These had a mean length of  $13 \pm 5$  mm.<sup>54</sup> The authors concluded that urachal remnants >22.5 mm (95th percentile in their review) should be considered abnormal findings. Although subtle, umbilical retraction with voiding (in association with pain) can be a sign of urachal anomalies.<sup>55</sup> Urinary ascities in the neonate due to perforation of the urachus at the time of umbilical artery/vein catheterization have been reported.<sup>56</sup> Many children with urachal cysts present shortly before or during adolescence, with inflammatory changes mimicking appendicitis. In this setting, a CT scan may suggest the diagnosis.<sup>57</sup> Some urachal abnormalities may not present until adulthood. In one study of 41 patients with urachal abnormalities treated at the Ochsner clinic, approximately one-fourth (11/41) were adults.<sup>58</sup>

## Infection

Infected urachal cysts usually present as possible appendicitis in the older child or young adult. They may be an

unsuspected finding at operation. An infected urachal cyst is traditionally managed in two stages: initially with antibiotics and drainage (either surgical or via interventional radiology), followed by delayed resection once the infection has resolved.<sup>59-61</sup> The latter procedure can be done either open or via laparoscopy.<sup>62-64</sup> However, there are some concerns regarding incomplete resection with laparoscopy. Gram-positive organisms are usually causative (Staphylococci and E. Coli are most common).

## Patent Urachus

Much of the urachal literature consists of case reports, rendering sound conclusions difficult. There have been rare reports of a patent urachus closing in the early newborn period.<sup>65,66</sup> A patent urachus can result in bladder prolapse,<sup>67,68</sup> or a “giant” umbilical cord.<sup>69</sup> Patency of the urachus is estimated to account for about 10% to 15% of urachal anomalies.<sup>65</sup> A patent urachus may sometimes present as a pseudocyst of the cord in the antenatal/fetal period.<sup>70</sup>

## Treatment

Excision is the treatment of choice for urachal abnormalities. A curvilinear infraumbilical incision (infants) or a transverse incision midway between the umbilicus in pubis (in older children) provides adequate exposure. The tract/cyst and a small cuff of bladder at the insertion are removed. Mucosa should not be left at the umbilicus. As mentioned above, laparoscopic resection has been used successfully as well.

## Malignancy

Standard treatment of urachal remnants includes resection of a cuff on urinary bladder at the entry site of the urachus. The histology of the excised bladder mucosa has been examined.<sup>71</sup> In one study, 17/23 patients undergoing resection of urachal remnants had a normal epithelial lining, and 25% (6/23) had abnormal bladder epithelium. The authors concluded that excision of a cuff of bladder was indicated. The risk of future cancer in urachal remnants is well recognized. Urachal cancers account for only 1% to 10% of adult bladder cancers, with a 10-year disease-free survival of about 50%.<sup>72,73</sup> Although malignancy related to the urachus usually develops in late adulthood, there have even been reports in adolescence.<sup>74</sup>

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