

# Associated patent urachus and patent omphalo-mesenteric duct in children: review of the literature

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The objective of this article is to review the literature on a very rare association such as patent urachus and a patent omphalo-mesenteric duct in children. Partial or total failure of the obliteration of urachus gives rise to various anomalies, which can be diagnosed both in childhood and in adulthood. The omphalo-mesenteric duct remnants are the most common anomalies of the gastro-intestinal tract, often asymptomatic. The association of patent urachus and a patent omphalo-mesenteric duct in children is very rare. A computer-assisted (PubMed and Google Scholar) search of the pediatric literature to identify all cases of patent urachus and a patent omphalo-mesenteric duct association was performed. We found only eleven cases of the association of patent urachus and a patent omphalo-mesenteric duct in the pediatric literature. All cases except one were males. When reported, all children affected were full term. The clinical sign reported was umbilical spillage. In all cases was reported a lack of healing of the umbilical stump with different clinical pictures, except two cases. Treatments reported were application of silver nitrate, antibiotic cream, povidone iodine, and application of salves and plasters. Various diagnostic examinations were

performed. In none of these patients was the possible association with PU and POMD suspected, but it was evidenced only during the surgical excision. The surgical approach was laparotomy in all cases. The association of patent urachus and a patent omphalo-mesenteric duct in children is very rare approached in all cases by laparotomy. This review underlines the importance of evaluating any persisting umbilical lesions without delay when conventional pharmacological therapy fails. *Ann Pediatr Surg* 13:113–115 © 2017 Annals of Pediatric Surgery.

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## Introduction

The urachus is an embryological remnant of the allantois. Between the fourth and the fifth month of gestation, the fetal bladder tends to descend gradually and its apical portion creates the urachus. The urachus obliterates itself, becoming the median umbilical ligament. Its persistence after intra-uterine life can manifest as different pathologies called urachal remnants (URs). A patent urachus (PU) is because of the complete lack of obliteration of the urachus.

The vitelline duct is the embryological narrow channel connecting the yolk sac with the intestine. During the fetal life, the vitelline duct tends to obliterate itself till to disappear. The partial failure of this obliteration can lead to the development of a duct, a ligament, or a diverticulum known as Meckel's diverticulum. A patent omphalo-mesenteric duct (POMD) occurs because of the complete lack of obliteration of the vitelline duct. The association of both anomalies is very rare.

## Methods

A computer-assisted (PubMed and Google Scholar) search of the pediatric literature to identify all cases of PU and POMD association was performed to evidence the different clinical management and surgical approaches. This review has been performed according to the ethical standards of the Committee on Human Experimentation of the institution in which the experiments were conducted or in accord with the ethical standards of the Helsinki Declaration of 1975.

## Results

We found only eleven cases of the association of a PU and a POMD in the pediatric literature from 1898 to 2016 (Table 1). All cases except one were males. When reported (5/11), all children affected by this association were full term. Specific signs reported were the spillage of different materials such as urine, mucous, soft stools, and clear fluid. Signs at physical examination were in all cases a lack of healing of the umbilical stump with different clinical pictures, except in two cases: one with an evident fistula and one with bowel prolapse through the umbilicus. The first attempt at treatment was reported in four cases and consisted in one case of local application of silver nitrate, and in three single cases of application of silver nitrate along with an antibiotic cream, povidone iodine, and application of salves and plasters. Diagnostic examination was performed with IV Indigo Carmine in one patient, cystogram, fistulography, and magnetic resonance in another case, and ultrasonography (US) and cystogram in another case. In none of these patients was the possible association with PU and POMD suspected, but it was evidenced only during the surgical excision. The surgical approach was by laparotomy in all cases, in four cases by a subumbilical transverse incision, in three cases by an elliptic incision about the umbilicus, in one case by an umbilical circumcision, and in another case by a Pfannenstiel incision. In two cases, the approach was not reported. When reported, the final diagnosis was made only intraoperatively and incidentally in eight cases, in one case only after histopathological examination, and in another case, the intraoperative diagnosis confirmed the preoperative suspicion.

**Table 1 Cases of associated patent urachus and patent omphalo–mesenteric duct reported in the literature**

Patient nos	References	Age	Sex	Gestational age	First attempt treatment	Signs at physical examination	Specific signs	Examinations	Preoperative diagnosis	Surgical approach	Diagnosis of PU associated with POMD
1	Lexer 1	1.5 years	M		Salves and plasters	Umbilical fistula in previous umbilical reddish tumor	Umbilical spillage of urine	None	PU		Histopathological
2	Cullen 2	14 months	M			Umbilical bleeding	Umbilical spillage of feces and urine			Elliptic incision about the umbilicus	Incidental intraoperative
3	Davis and Niehaus 3	1 year	M		Silver nitrate	Elevated umbilical pouch covered with red mucous membrane and bled easily	Umbilical spillage of feces and urine	None		Elliptic incision about the umbilicus	Incidental intraoperative after bleeding of cut of omphalo–mesenteric artery
4	Nerdrum 4	17 days	M	Full term		Nonhealing navel flesh-colored tumor protruding from the umbilical opening	Umbilical granuloma	None	Omphalocele/ OMD	Umbilical circumcision	Incidental intraoperative
5	Griffith <i>et al.</i> 5	1 month	M	Full term		Umbilical granuloma	Umbilical spillage of liquid, mucus, and soft stools	None		Elliptic incision about the umbilicus	Incidental intraoperative
6	Alessandrini and Derlon 6	–	M				–	None			
7	Fujiwara <i>et al.</i> 7	8 months	M	Full term	Povidone iodine	Umbilical granuloma	Umbilical spillage of mucus and yellow liquid	IV Indigo Carmine	PU	Pfannenstiel incision	Incidental intraoperative
8	Lizerbram <i>et al.</i> 8	6 days	M				Umbilical spillage of yellow fluid	Cystogram Fistulography MR	Suspected association of PU and POMD	Sub-umbilical transverse incision	Confirmed intraoperatively
9	Sharma <i>et al.</i> 9	21 days	F	Full term		intestine prolapsing through the umbilicus	Previous reported spillage of feces and clear fluids	None		Subumbilical transverse incision	Incidental intraoperative
10	Chawada and Ghavghave 10	6 weeks	M	Full term		Swelling on the umbilical region	Previous reported spillage of feces and clear fluids	None	POMD	Subumbilical transverse incision	Incidental intraoperative
11	Gupta <i>et al.</i> 11	2 months	M		Antibiotic cream and silver nitrate	umbilical induration	Serous umbilical spillage	US, cystogram	Urachal anomaly	Sub-umbilical transverse incision	Incidental intraoperative

F, female; M, male; MR, magnetic resonance; OMD, omphalo–mesenteric duct; POMD, patent omphalo–mesenteric duct; PU, patent urachus; US, ultrasound.

## Discussion

Partial or total failure of the obliteration of urachus gives rise to various anomalies, which can be diagnosed both in childhood than in adulthood. The incidence of these anomalies was one to two in 10 000 deliveries [12]. URs can manifest as PU, urachal cysts, urachal sinus, and bladder diverticulum.

Omphalo–mesenteric duct remnants are the most common anomalies of the gastrointestinal tract, often symptomatic [13]. These anomalies may range from POMD to the most common Meckel's diverticulum.

Omphalo–mesenteric duct malformations may become symptomatic at any age [14] and common symptoms include abdominal pain, intestinal bleeding, intestinal obstruction, umbilical drainage, and umbilical hernia. Infections and tumors are the two main complications of urachal anomalies: in the first case, the drainage of infectious fluids can take place in the bladder, umbilicus, or both [15].

Urachal tumors may either be benign or malignant. Urachal carcinomas are typically silent because of their extraperitoneal location and therefore most of the patients present at diagnosis local invasion and/or metastasis detected by US and computed tomography.

Patients without epithelialization of URs appear to have a low risk of malignant transformation. Copp *et al.* [16] analyzed the specimens of 29 URs excised in children to predict a possible presence of epithelium before the removal. Unfortunately, they concluded that it is not possible to predict which patients have epithelialized URs with only radiological images or symptomatology.

Nowadays, as a first attempt, the diagnosis of URs is made on the basis of US. Nevertheless, computed tomography is often the preferred method for a definitive diagnosis as reported by Widni *et al.* [17], affirming that the US in the diagnosis of URs lacks accuracy in distinguishing true negatives from false positives.

Nevertheless, in our experience [12,14], US plays an important diagnostic role in this kind of anomalies if performed by expert physicians and, in our opinion, this diagnostic tool could receive a wider application.

In the literature, there are many studies on the diagnosis and treatment of URs and omphalo-mesenteric duct remnants. Nevertheless, the association of a PU and a POMD in children is rarely reported. In our review of the pediatric literature, we found only eleven cases [1–11] from 1898 to 2016.

All cases except one [4] were males, with a M:F ratio of 10:1. It seems that this association is not correlated to prematurity because when reported (6/11), all affected newborns were full term. Specific signs reported were the spillage of different materials such as urine, mucous, soft stools, and clear fluid. Signs at physical examination were in all cases a nonhealing navel reported in different manners, except in two cases: one with an evident fistula and one with bowel prolapse through the umbilicus. Nevertheless, in none of these patients was the possible association with PU and POMD suspected, but was only evidenced during the surgical excision. The first attempt at treatment was reported in five cases and consisted in two cases of local application of silver nitrate, and in three single cases of silver nitrate along with an antibiotic cream, povidone iodine, and application of salves and plasters. All these attempts failed to resolve the problem of a nonhealing navel.

Even though the modern approach to urachal anomalies is US, this tool was used only in one case. In fact, a diagnostic examination was performed with IV Indigo Carmine in one patient, cystogram, fistulography, and magnetic resonance in another case, and US and cystogram in another one. In none of these patients was the possible association with PU and POMD diagnosed, but was only evidenced during the surgical excision.

Recent reports advocate laparoscopy for the excision of urachal anomalies and also for Meckel's diverticulectomy, but in the case of associated PU and POMD [18,19], our review showed only laparotomic approaches.

This surgical approach was never used to make the right diagnosis or to excise the remnants and, in all cases, the patients were approached by laparotomy, in four cases by a subumbilical transverse incision, in three cases by an elliptic incision about the umbilicus, in one case by an umbilical circumcission, and in another case by a Pfannenstiel incision. In two cases, the approach was not reported. When reported, the final diagnosis was made only intraoperatively and incidentally in eight cases, in one case only after histopathological examination, and in another case, the intraoperative diagnosis confirmed the preoperative suspicion.

The approach of a nonhealing navel in newborns has changed considerably over the past century, with a reduced focus on cystography and fistulography and more invasive methods compared with US. The improving resolution of US in the last decades has led to consider such exams as a first attempt to define the umbilical anomalies. The relationship between PU and POMD,

however, being extremely rare, is also difficult to evidence. From this point of view, the US approach should be aimed at evaluating both the UR and other possible associated malformations to better define the anatomy of the region before considering any surgical approach. Laparoscopy could confirm the suspicion and, being a mini-invasive technique, could be considered a possible future gold-standard approach to avoid further invasive diagnostic exams with the advantage of making the correct diagnosis and the possibility to proceed to the excision of both remnants with minimal incisions.

Our review underlines the rarity of this association that was never diagnosed preoperatively. Furthermore, it indicates the importance of evaluating any persisting umbilical lesions without delay when conventional pharmacological therapy fails. However, in case of fecal or urinary discharge, a fast instrumental diagnosis and surgical intervention are justified without even attempting medical therapy.

### Conflicts of interest

There are no conflicts of interest.

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