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## Giant umbilical cord



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A giant umbilical cord is a very rare malformation of the umbilical cord that can easily be diagnosed on prenatal scans and is unmistakable postnatally. We present a typical case to demonstrate aspects of this rare diagnosis relevant to obstetricians, neonatologists and also parents of patients with this malformation.

In a 37-year-old G2/P1, with an unremarkable medical and obstetric history, routine ultrasound at 20 weeks of gestation revealed a live singleton fetus with two anechoic umbilical cystic masses (measuring 30 and 20 mm in diameter) at the fetal insertion site (Fig. 1A). Ultrasonographic color Doppler imaging confirmed normal flow through two umbilical arteries and one umbilical vein (Fig. 1B; movie). The vessels were clearly separate from the cystic masses. The umbilical cord insertion on the fetal abdomen appeared normal and no other anomalies were noted. A connection between the cysts and the fetal bladder was not apparent (Fig. 1B). The amniotic fluid index was normal.

Serial US examinations at 28, 32, 34 weeks gestation documented appropriate fetal development. The size and the number of the cysts increased. On the last US performed at 37 weeks gestation, five large umbilical cysts were apparent, measuring between 30 and 70 mm in diameter. An emergency Cesarean section was performed at 39 weeks of gestation because of re-

## INTRODUCTION

## CASE REPORT

peated fetal bradycardia after spontaneous labor had started. Membranes were still intact and amniotic fluid was clear.

An SGA male infant with a birth weight of 2800 g (< P10), length of 48 cm (< P10) and head circumference of 33.5 cm (P10) was easily extracted and a giant umbilical cord was clamped approximately 30 cm from the abdominal wall, where it became thinner (Fig. 2). Neonatal adaptation was excellent with Apgar scores of 9, 10, 10 at 1, 5 and 10 minutes, respectively. Umbilical cord pH of 7.25 was available from one vessel only.

On clinical examination, the cutaneous umbilicus was very large (3 cm in diameter) and the lobulated gelatinous part measured 28 x 12 cm. An enormous, homogeneous, transparent and fluctuating jelly appeared as hypertrophic Wharton's jelly and allowed easy recognition of the three umbilical vessels. At the center of the emerging gelatinous umbilicus a thin reddish structure was noted and was interpreted as a localized hemorrhage or neovascularization.

An US examination of the base of the umbilicus, performed to exclude a patent urachus before clamping the cord more proximally, was inconclusive. Therefore, it was decided to clamp the cord just above the skin and to send the stump for histological examination (Fig. 3). Kidneys were normal on ultrasound.

Subsequently, the newborn did well and the dried umbilical stump detached after ten days, leaving a granulomatous structure. Since the umbilicus was dry, the child returned home with instructions for cord care. One week later, the boy presented again to the hospital because of discharge of transparent fluid from the umbilicus, noted by the mother during urination and coughing (Fig. 4). At this time, US examination revealed a probable communication with the bladder (Fig. 5), which was confirmed by cysto-urography (Fig. 6). Further pathologies of the urinary tract such as posterior urethral valves or vesico-ureteral reflux were excluded. Surgical resection of the persistent patent urachus was performed at 19 days of life (Fig. 7) followed by an uneventful postoperative course.

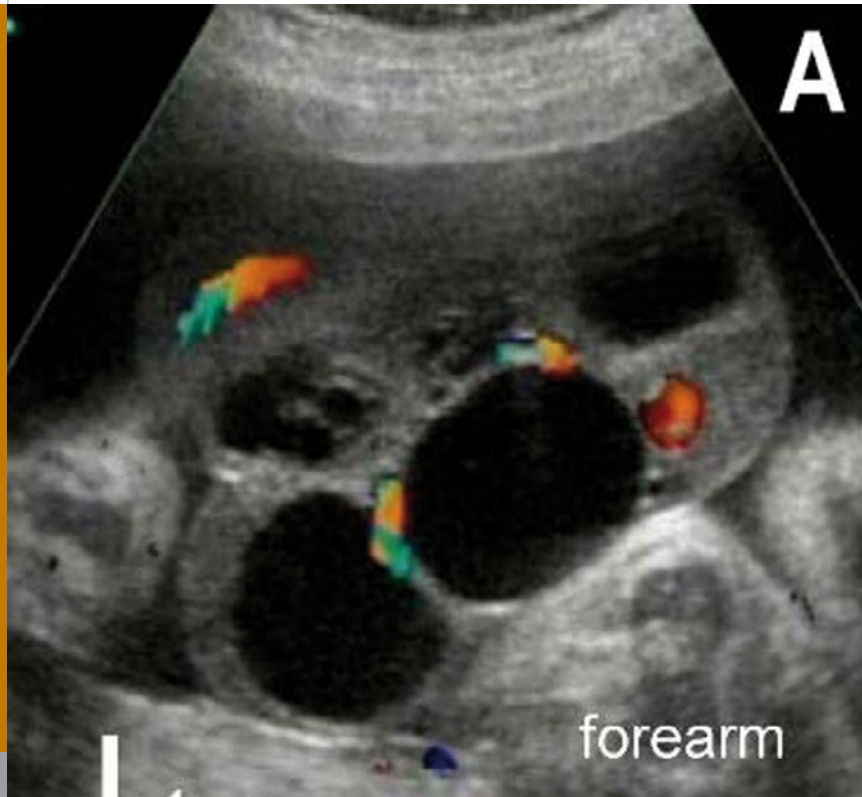


Fig. 1A

*Prenatal US scan: A) View of the umbilical cord with several large cysts in Wharton's jelly.*

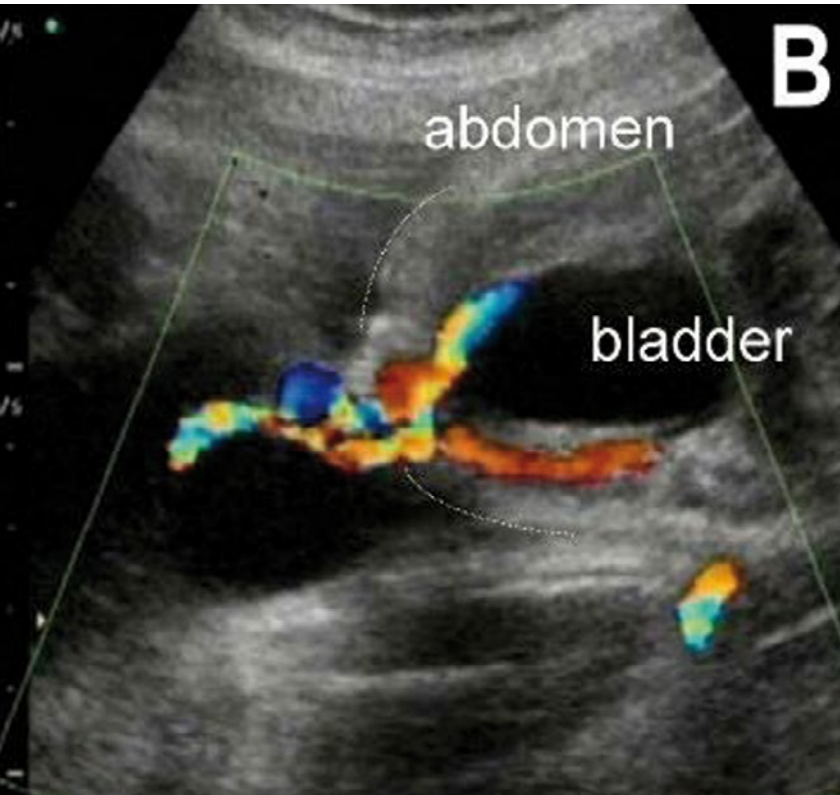


Fig. 1B

*Prenatal US scan: B) Section of abdomen with bladder and umbilical vessels.*



Fig. 2

*Postnatal appearance of the umbilical cord: large, lobulated and transparent umbilical cord with unusually large cutaneous rim at the base.*





Fig. 3

*Presence of the allantois duct and multilocular cysts with severe pericyclic edema. Inserts show magnification of allantois duct, with hematoxylin-eosin (HE) and keratin immunostaining (Kerat). The central conduit confirms patent urachus.*



**Fig. 4**

*Discharge of urine from the umbilicus.*

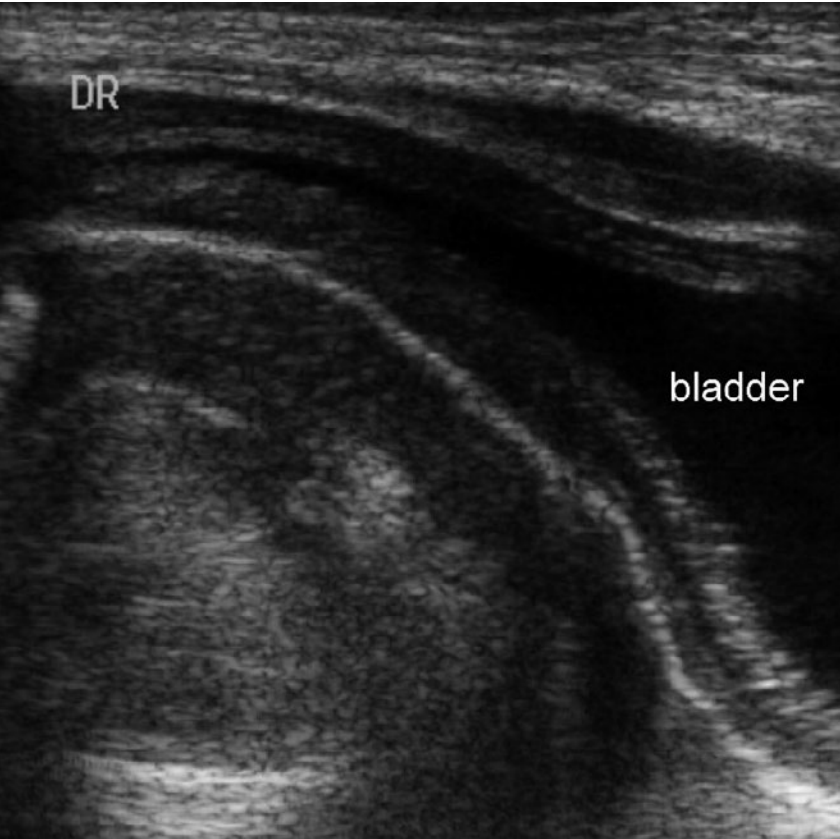
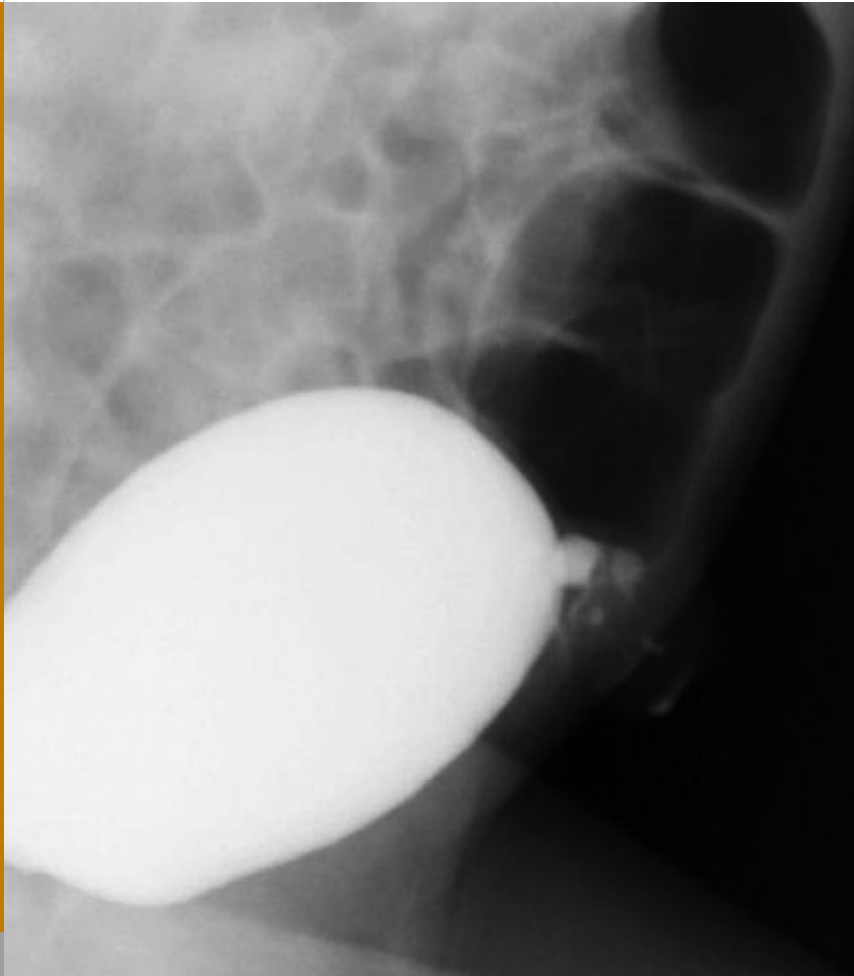


Fig. 5

*Bladder ultrasound. Presence of a narrow conduit between the roof of the bladder and the umbilicus suggesting patent urachus.*



**Fig. 6**

*Cysto-uretrography. Presence of a patent conduit between the anterior upper pole of the bladder and the umbilicus, contrast is exiting through the umbilical stump.*

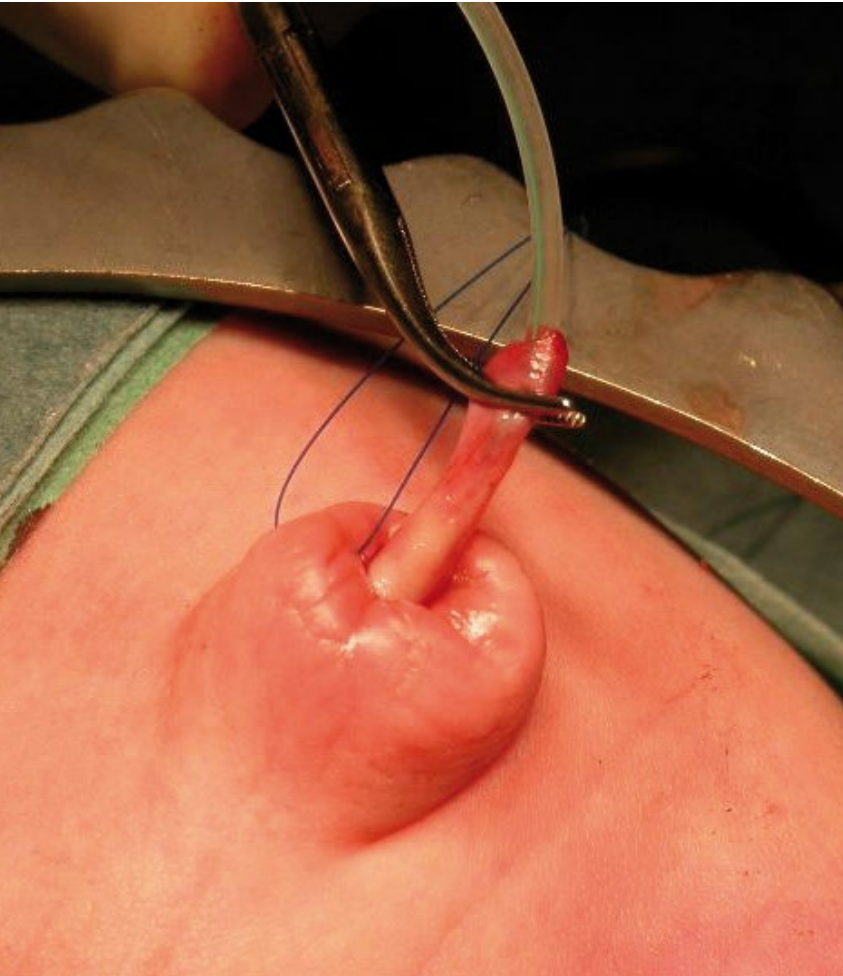


Fig. 7

*Dissected patent urachus everted over the catheter.*

## DISCUSSION

Umbilical cord cysts develop from the partial or complete absence of obliteration of the allantois or omphalo-mesenteric duct. The prenatal differential diagnosis includes pseudocysts (degeneration of Wharton's jelly), omphalo-mesenteric duct cysts, vascular disorders, abdominal wall defects, bladder exstrophy and urachal anomalies (3).

Congenital patent urachus is an extremely rare condition with an incidence of 1-2.5:100'000 deliveries (1, 3, 4). Males are affected twice as often as females (1). On prenatal US, the presence of a direct communication between the fetal bladder and the umbilical cord cyst confirms the diagnosis of a patent urachus (1, 5). This pathognomonic feature, however, is not always apparent prenatally (4). Congenital patent urachus is rarely associated with other anomalies such as posterior urethral valves, and, when isolated, not related to chromosomal defects.

When detected prenatally, congenital patent urachus should lead to close monitoring and follow-up of the mother and fetus (3) because of a possible compression effect of the cystic mass on the umbilical vessels, particularly at term and during labor, resulting in fetal compromise as in the present case. The preferred mode of delivery may be discussed based on considerations the size of the lesion (4).

The general use of plastic clamps on the umbilical stump makes detection of the condition often difficult for the pediatrician during neonatal evaluation. The giant umbilical cord is a very rare finding and only very small case series have been published (6). As its development is likely to result from liquid flowing into the Wharton's jelly, we now would recommend a thorough investigation of the umbilical stump shortly after birth, including US, cysto-urography and histology of the stump. Most causes of giant umbilical cords will require surgical intervention (2).

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