

## Short Communication

# Congenital appendicoumbilical fistula

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## 1. Introduction

Congenital umbilical anomalies are common in children, and the most prevalent types are persistent urachal remnant and persistent omphalomesenteric duct (POMD). The usual clinical presentation is that of an umbilical granuloma or failure of umbilical artery catheterization. We report a very rare case of congenital appendicoumbilical fistula (AUF), provide an overview of this diagnosis, and discuss the etiology and management of the condition in the context of this patient with AUF. We further present a review of the literature.

## 2. Case report

A full-term, 2-month-old girl with no personal medical history and no family history was referred with an umbilical granuloma that had been treated unsuccessfully with silver nitrate. No anomaly was detected in prenatal examinations, and no attempt had been made to catheterize the umbilical vessels. She presented normal height and weight for age and normal bowel function. A red mass protruded from the umbilicus, with a central orifice producing serous discharge (Supplementary figure 1). Neither urinary nor fecal discharge was observed. The opening was easily catheterized. Persistent urachus was suspected but was not confirmed by ultrasound. Surgical exploration was therefore indicated.

The operative procedure was performed via an umbilical incision. Interestingly, the granuloma was found to correspond to the tip of the vermiform appendix normally inserted into the cecum (Fig. 1). No signs of malrotation were observed. There was no significant urachal remnant or Meckel's diverticulum. An appendectomy was performed. The patient was discharged home on postoperative day two. Her postoperative recovery was uneventful. Histopathology found a normal vermiform appendix.

## 3. Discussion

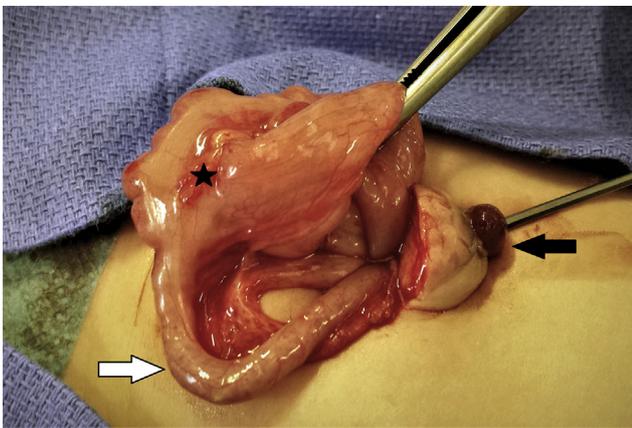
Umbilical malformations are common in children. This case of AUF presented as an umbilical granuloma. AUF, also known as umbilical appendix, is rare: fewer than 15 cases have been reported in the literature since 1921.<sup>1</sup>

The origin of AUF remains unclear. Two theories have been proposed in the literature. According to the first theory, the vermiform appendix herniates through the umbilicus due to a mobile cecum. The umbilical location of the appendix is due to either failure to re-enter the peritoneum from the extraembryonic coelom or secondary herniation of the appendix through the umbilical ring, related to a mobile cecum. According to the second theory, AUF is suspected to be a POMD arising from appendix. A POMD arising from the vermiform appendix may generate an umbilical hernia, into which the tip of the appendix drains.

Despite insufficient evidence as to the cause of AUF, the most likely hypothesis is that ligation of the umbilical cord containing the tip of the appendix, causing injury to the

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**Figure 1** Intraoperative view. Tip of the vermiform appendix draining into the umbilicus. *Black arrow:* Umbilical granuloma. A catheter is inserted into the appendix through the umbilical granuloma. *White arrow:* Vermiform appendix. *Black star:* Cecum.

appendix, is responsible for the condition. In one case report,<sup>2</sup> the vermiform appendix was entirely located in an umbilical hernia. As the vermiform appendix is typically described as being the last part of the gut to re-enter the peritoneal cavity, failure of re-entry could explain umbilical herniation and formation of an AUF due to injury of the appendix caused by umbilical cord clamping (Supplementary figure 2).

No risk factors for AUF have been identified due to the small number of cases reported in the literature. No malformation was associated with AUF in previously published cases. Nevertheless, AUF is suspected to be associated with a certain degree of malrotation. A mobile cecum,<sup>1</sup> which cannot be considered as malrotation per se, could facilitate herniation of the appendix into the umbilical cord, especially when it is located close to the midline.

An umbilical mass is considered abnormal when it persists after the age of 2 months despite adequate medical treatment.<sup>3,4</sup> The first-line treatment for an umbilical granuloma is silver nitrate therapy, which can be started once the umbilical stump has been shed.<sup>5</sup> Between two and three local applications can be attempted. Persistence of an umbilical granuloma despite a well-conducted medical treatment excludes the diagnosis of simple granulation tissue and constitutes an indication for further exploration. Observation of a central orifice producing urinary, fecal, or bilious discharge is immediately considered abnormal and renders unnecessary any local treatment. Imaging serves as a tool for preparing the surgical procedure, seeking a differential diagnosis, and providing information to the patient's family. Ultrasonography is the initial screening test, used to search for a longitudinal double line from the umbilicus to either the midgut in the case of POMD or the bladder in the case of a urachal remnant.<sup>6</sup> Plain abdominal radiography is normal. A computed tomography (CT) scan can aid in the diagnosis but, due to the radiation exposure involved, is not the first choice. The accuracy of CT for urachal remnants depends on the bladder's filling status.<sup>7</sup> Magnetic resonance imaging can also be considered. The

administration of CT and magnetic resonance imaging for the diagnosis of umbilical fistula is rarely described in the literature. A connection between the umbilical anomaly and the digestive tract is suggestive of POMD.

The value of fistulography is important in the exploration of an umbilical abnormality. Fistulography showing contrast passing into the bowel suggests the existence of a POMD. As all patients require surgical exploration,<sup>5</sup> we do not recommend invasive imaging for a persistent umbilical granuloma in our center.

Surgery is required in all cases of persistent granuloma producing discharge. The operation can be performed as soon as the umbilical mass is considered abnormal. Surgery accomplishes the final diagnosis and treatment of the anomaly. An appendectomy was performed in all published cases of AUF, and normal histology was found in every case. No surgical complications have been reported.

#### 4. Conclusion

Umbilical appendix is an incidental finding of uncertain origin. AUF is probably the result of injury to the tip of the appendix in an unrecognized umbilical hernia caused by neonatal umbilical cord clamping. The congenital nature of AUF therefore cannot be confirmed definitively in every case. Surgical exploration is indicated.

#### Conflicts of interest

None.

#### Acknowledgments

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#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.pedneo.2019.09.009>.