Scientific Research Publishing

Surgical Science, 2026, 17(3), 129-136
https://www.scirp.org/journal/ss
ISSN Online: 2157-9415
ISSN Print: 2157-9407

# Rare Association of Congenital Megaoesophagus and Aortomesenteric Clamp Syndrome (AMCS): An Exceptional Case in an Infant at Yaoundé Central Hospital (YCH)

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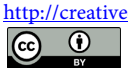
**How to cite this paper:** Dorcas, N.B., Antoine, B.S., Francis, N., Patrick, B.B.D.C., Laetitia, M., Eric, N., Roger, B.M.G., Stéphane, E.M.E., Valery, M.P., Jessica, D.J., Raïssa, M.B.T., Diana, E., Amenguele, B., René, N. and Félicien, M.T.F. (2026) Rare Association of Congenital Megaoesophagus and Aortomesenteric Clamp Syndrome (AMCS): An Exceptional Case in an Infant at Yaoundé Central Hospital (YCH). *Surgical Science*, **17**, 129-136.
https://doi.org/10.4236/ss.2026.173014

**Received:** February 24, 2026
**Accepted:** March 28, 2026
**Published:** March 31, 2026

## Abstract

Oesophageal achalasia is a rare oesophageal motility disorder in children and remains exceptional in infants. Its association with Superior Mesenteric Artery Syndrome (SMAS), characterized by compression of the third portion of the duodenum between the abdominal aorta and the superior mesenteric artery, is extremely uncommon and has mainly been reported in adults. We report the first case of this association, which occurred in a 10-month-old female infant presenting with chronic regurgitation since birth and severe acute malnutrition. Imaging investigations, including abdominal computed tomography and upper gastrointestinal contrast study, confirmed the diagnosis of oesophageal achalasia associated with SMAS. The patient underwent Heller cardiomyotomy combined with Dor fundoplication. In addition, during follow-up, further oesophageal dilatation using Savary-Gilliard dilators was done, resulting in progressive clinical improvement and weight gain. This case highlights the importance of considering SMAS in infants presenting with severe malnutrition secondary to achalasia.





## Keywords

Megaoesophagus, Achalasia, SMAS, Infant, YCH

## 1. Introduction

Megaesophagus or achalasia is a rare oesophageal motility disorder characterized by impaired relaxation of the lower oesophageal sphincter and absence of normal oesophageal peristalsis [1]-[3]. Although widely described in adults, the disease remains uncommon in the paediatric population and exceptional in infants [2] [3]. The estimated incidence in children is approximately 1 case per 100,000 births per year, and diagnosis is often delayed because symptoms such as regurgitation, vomiting or feeding difficulties are nonspecific [3] [4].

Superior Mesenteric Artery Syndrome (SMAS), also known as Wilkie syndrome, results from compression of the third portion of the duodenum between the abdominal aorta and the superior mesenteric artery [2] [5] [6]. Radiological diagnosis is suggested when the aorto-mesenteric angle is less than 25° and the aorto-mesenteric distance is less than 8 - 10 mm [2] [5]. The reported incidence ranges between 0.013% and 0.3% in the general population [7]. SMAS is frequently associated with severe weight loss, which reduces the mesenteric fat cushion normally maintaining the angle between the aorta and the superior mesenteric artery [5] [8]. Young female patients appear to be more predisposed to this condition [2] [5].

Although achalasia and SMAS are well described individually, their association is extremely rare and has mainly been reported in adults [1] [2]. Severe malnutrition caused by chronic achalasia may predispose patients to SMAS by reducing the protective fat pad surrounding the duodenum. Reports of this association in children remain exceptional and, to our knowledge, have rarely been described in infants.

We therefore report this inaugural case of congenital achalasia associated with SMAS in a 10-month-old infant managed at Yaoundé Central Hospital.

## 2. Case Presentation

A 10-month-old female infant was referred to our paediatric surgery department with suspected hypertrophic pyloric stenosis because of persistent non-bilious whitish vomiting evolving for two weeks and associated with significant weight loss and asthenia. Her medical history revealed chronic regurgitation since birth. Family history showed a congenital malformation (omphalocele) in an older sibling.

Clinical examination revealed severe acute malnutrition with deterioration of the general condition. The patient had lost approximately 4 kg in two weeks, corresponding to 29% weight loss. Mid-upper arm circumference measured 9 cm and anthropometric parameters were below −3 Z-score (**Figure 1**). This was associated with an occlusive syndrome of the upper digestive tract.

The diagnostic discussion included obstructive digestive pathology located between the oesophagus and the duodenum upstream of the ampulla of Vater. Differential diagnoses considered were congenital megaoesophagus, duodenal obstruction at the level of his first or second portion, digestive duplication of the supraduodenal organs, SMAS and complicated gastro-oesophageal reflux.

Abdominal computed tomography revealed dilation of the thoracic oesophagus and compression of the third portion of the duodenum between the abdominal aorta and the superior mesenteric artery. The aorto-mesenteric angle measured 17˚ and the aorto-mesenteric distance was 3 mm, values below the diagnostic thresholds reported for SMAS in the literature [2] [5] (**Figure 2**).

Upper gastrointestinal contrast study demonstrated significant oesophageal dilation with a typical "bird-beak" appearance and tortuosity of the distal oesophagus consistent with dolicho-megaoesophagus classified as type II according to Forbes (**Figure 3**). Upper gastrointestinal endoscopy revealed marked oesophageal dilation with an impassable stenosis at the level of the cardia, confirming the diagnosis of achalasia (**Figure 4**).

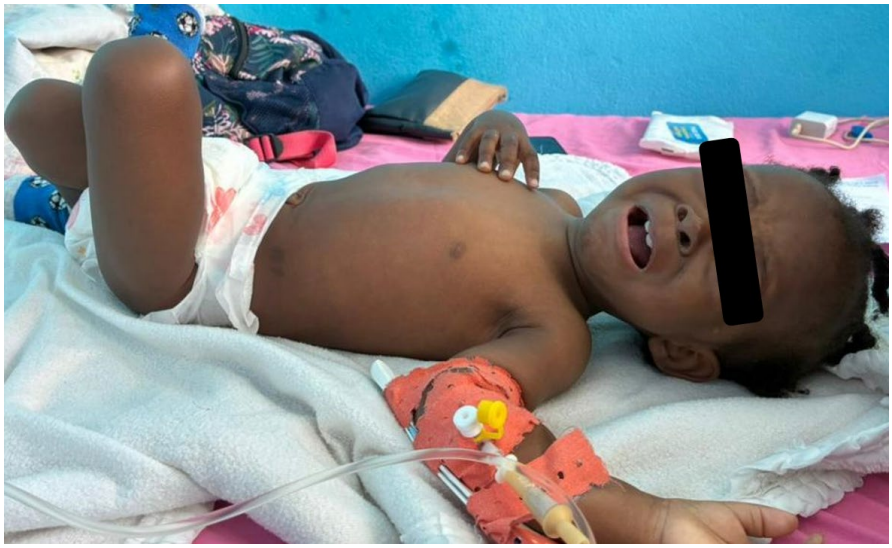


**Figure 1.** Image of the infant prior to surgery (YCH photo library).

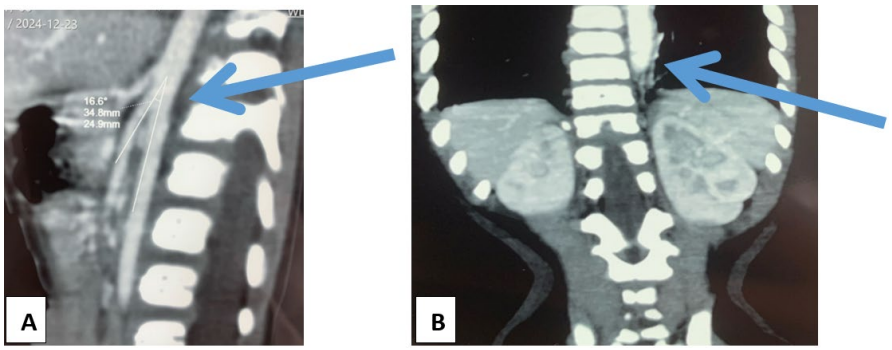



**Figure 2.** Abdominal scan images. (A) Aorto-mesenteric angle at 17˚, (B) Dilatation of the thoracic oesophagus (Photothèque YCH).

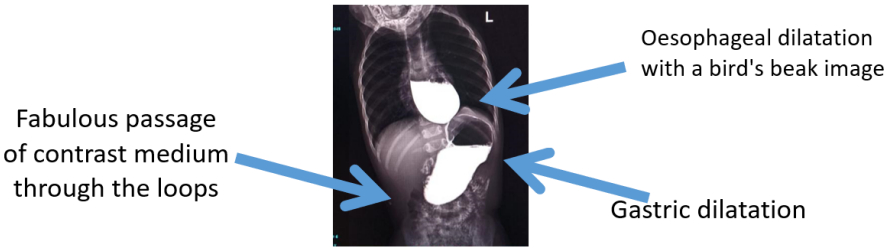



**Figure 3.** Preoperative upper gastrointestinal contrast study image (YCH Photo Library).

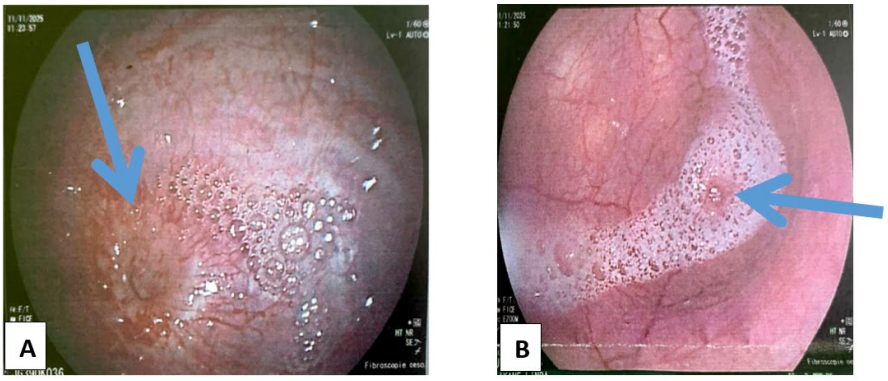



**Figure 4.** Esophagogastroduodenal fibroscopy images: (A) Oesophageal dilation; (B) Impassable stenosis of the cardia (YCH Photo Library).

Biological investigations revealed hypokalaemia (3.35 mmol/L) and anaemia (9.3 g/dL).

Achalasia was considered primary because no secondary causes such as neurological disease, metabolic disorders, infectious diseases or syndromic associations were identified during clinical and biological evaluation.

The final diagnosis was Forbes type 2 primary oesophageal achalasia associated with secondary SMAS complicated by severe acute malnutrition.

## 3. Therapeutic Management

### 3.1. Preoperative Management

Initial management included gastric decompression using a nasogastric tube and strict fasting. Parenteral rehydration was initiated with correction of electrolyte disturbances, particularly hypokalaemia. Blood transfusion was performed to correct anaemia. Nutritional support with Astymin® was also introduced to improve the patient's general condition before surgery.

After approximately ten days of medical stabilization, the patient gained about 1 kg and surgical intervention was performed.

### 3.2. Surgical Treatment

The surgical procedure consisted of Heller cardiomyotomy combined with Dor fundoplication (Figure 5). The distal oesophagus was mobilized and an extra-mucosal myotomy was performed on the anterior wall of the oesophagus extending

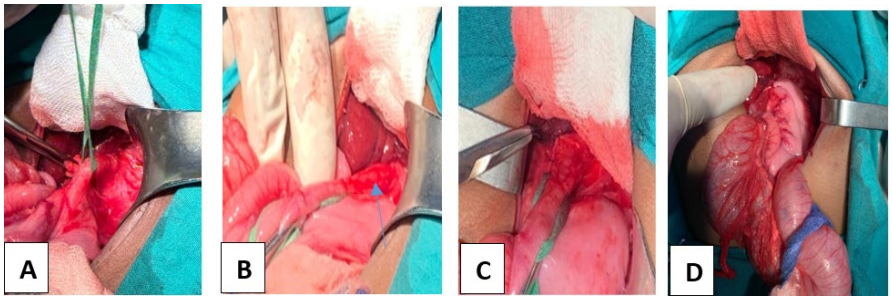



**Figure 5.** Heller's cardiomyotomy + Dor's fundoplication. (A) Mobilisation of the abdominal oesophagus. (B) Extra-mucosal myotomy. (C) Anchoring of the serosa to the pillars of the diaphragm. (D) Dor's fundoplication. (YCH Photo Library).

approximately 5 cm proximally and about 2 cm onto the gastric cardia to ensure complete division of the circular muscle fibres while preserving the mucosa. Anterior Dor fundoplication was then performed to protect the exposed mucosa and prevent postoperative gastro-oesophageal reflux.

Given the secondary nature of SMAS related to severe malnutrition, no surgical treatment was performed for duodenal compression, with the expectation of spontaneous resolution after nutritional recovery.

### 3.3. Postoperative Course

Postoperative management included analgesics, antibiotic therapy and proton pump inhibitor treatment. Enteral feeding through the nasogastric tube was initiated on the second postoperative day with careful clinical monitoring (digestive tolerance, resumption of bowel movements, weight).

Local care included a dressing applied on the fifth postoperative day.

### 3.4. Progress and Monitoring

Kinking of the NG tube led to its removal on the second postoperative day. Gradual oral feeding was started on the fourth day, initially with small amounts of water, then gradually with milk and enriched porridge.

The resumption of bowel movements was noted on the sixth day with greenish stools. However, regurgitation gradually reappeared, leading to increased meal frequency. Discharge was authorised on the tenth postoperative day with a weight gain of 700 g.

However, follow-up upper gastrointestinal contrast study performed two weeks later revealed residual oesophageal stenosis (**Figure 6**) associated with persistent regurgitation despite an additional weight gain of 300 g. This warranting further education for the parents, particularly regarding the semi-sitting position during and after meals and the administration of small portions of food.

At one-month follow-up, weight remained stable at 6.85 kg with persistent regurgitation. Endoscopic oesophageal dilatation using Savary-Gilliard dilators was performed with the insertion of a feeding tube (**Figure 7**), allowing administration of high-calorie nutritional formula (F100). Seven days after dilation, the patient's

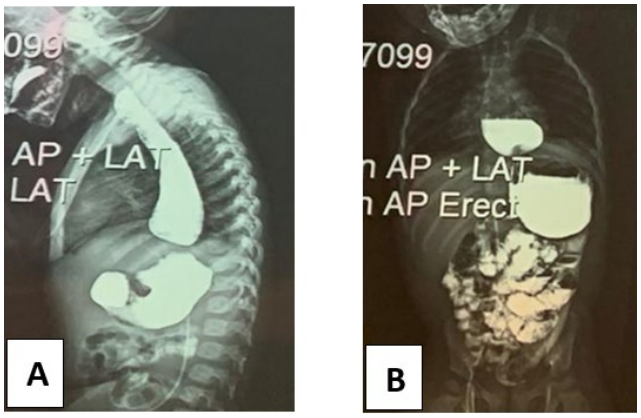



**Figure 6.** Upper GI contrast study control images. (A) Residual oesophageal stenosis. (B) contrast agent passing through the loops (YCH Photo Library).

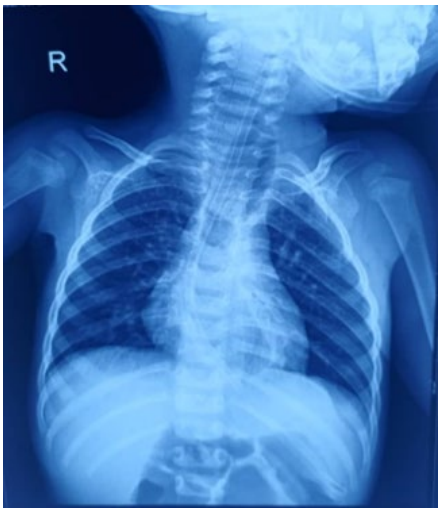



**Figure 7.** X-ray after oesophageal dilatation and insertion of a nasogastric tube (YCH photo library).

weight increased to 7.5 kg with significant clinical improvement.

### 3.5. Clinical Timeline

The clinical course of the patient can be summarized as follows: chronic regurgitation had been present since birth. At the age of 10 months, symptoms worsened with persistent vomiting and severe weight loss leading to hospital admission. After correction of hydro-electrolytic disorders and nutritional stabilization, surgical treatment was performed on the tenth day of hospitalization. Persistent postoperative symptoms led to oesophageal dilatation one month later, resulting in progressive weight gain and improved feeding tolerance.

## 4. Discussion

Achalasia remains a rare disease in childhood and is exceptional in infants, which

explains the frequent delay in diagnosis and the risk of initial misdiagnosis as more common conditions such as gastro-oesophageal reflux or hypertrophic pyloric stenosis [3] [4].

Diagnosis is usually suggested by imaging studies demonstrating oesophageal dilation with distal narrowing and the characteristic "bird-beak" appearance on contrast studies [9] [10]. High-resolution oesophageal manometry is considered the gold standard for diagnosis because it demonstrates impaired relaxation of the lower oesophageal sphincter and absence of oesophageal peristalsis [6] [11]. However, this investigation is not always available in low-resource settings, as in our case.

SMAS is an uncommon cause of upper intestinal obstruction resulting from compression of the third portion of the duodenum between the aorta and the superior mesenteric artery [6] [8]. The condition is often associated with severe weight loss leading to reduction of the mesenteric fat pad and narrowing of the aorto-mesenteric angle [5] [6]. In our patient, severe malnutrition caused by chronic achalasia likely contributed to the development of secondary SMAS.

Heller cardiomyotomy combined with Dor fundoplication remains the standard surgical treatment for achalasia in many centres, particularly in developing countries where advanced endoscopic techniques may not be available [4] [11] [12]. In infants, however, the procedure can be technically challenging due to small anatomical structures and poor nutritional status, which increase the risk of incomplete myotomy [4] [12].

Persistent symptoms after surgery may occur when the myotomy is incomplete, leading to residual oesophageal stenosis. In such cases, endoscopic oesophageal dilatation represents an effective, repeatable and minimally invasive therapeutic option [8] [10] [13]. Recent advances in the management of achalasia include laparoscopic Heller myotomy and peroral endoscopic myotomy (POEM), which have demonstrated excellent outcomes but remain unavailable in many low-resource settings [9] [11] [13].

The prognosis of SMAS secondary to malnutrition is generally favourable when nutritional rehabilitation restores the mesenteric fat pad and relieves duodenal compression [1] [14]-[16].

## 5. Conclusion

The association of congenital achalasia and Superior Mesenteric Artery Syndrome in infants is extremely rare. This case illustrates the central role of severe malnutrition caused by achalasia in the development of SMAS. Management should be comprehensive, prioritised, and directed toward treating the underlying condition. Heller's cardiomyotomy combined with Dor fundoplication, although technically demanding in infants, remains the standard treatment in our practice. In case of incomplete myotomy, secondary oesophagial dilatation may be required. Adequate nutritional management is essential and largely determines the prognosis of this association.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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