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Consideration of pyloric stenosis as a cause of feeding dysfunction in children with cyanotic heart disease

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Abstract

Feeding difficulty has been reported at a higher incidence in infants with cyanotic heart disease and single ventricle physiology necessitating specialized feeding strategies. However, structural causes of feed intolerance in this subset of patients should not be ignored. This case series highlights three recent cases of pyloric stenosis in infants with left-sided obstructive lesions at our institution. In all three cases, the initial presumed diagnosis was feeding intolerance related to heart disease, and there was significant clinical improvement following identification and correction of pyloric stenosis.

Keywords: Cyanotic heart disease, feeding, pyloric stenosis

INTRODUCTION

Feeding difficulties are common in patients with congenital heart disease. These include difficulty tolerating enteral feedings, longer time needed to reach full volume feeds, and additional need for specialized feeding therapies.[1] This is particularly the case in infants with cyanotic and complex heart disease such as hypoplastic left heart syndrome (HLHS). Data shows that these children have persistent feeding difficulties even after their initial cardiac surgery when compared to infants with other heart defects. Ultrasound measurements in a small group of infants with HLHS suggest that superior mesenteric artery perfusion is impaired both pre-Norwood and postoperatively, which may limit the body's ability to meet the increased metabolic requirements for feeding.[2] Risk of feeding difficulty is also higher in these patients because introduction of enteral feeds is often delayed due to the child's overall clinical state and complications related to cardiopulmonary bypass such as decreased bowel motility and pyloric dysfunction.[3] They have delays in feeding and oromotor readiness, gastric feeding, and development of oromotor skills.[4] An estimated 41%–48% of children with HLHS have feeding complications compared to only 4% of children with d-transposition of the great arteries (TGA) postcardiac surgery.[1,5] Davis *et al*. reported that 55% of the single-ventricle group required ancillary services to assist with feeding techniques as compared with 12.5% in the TGA group.[1] At our institution, approximately 65% of patients are discharged from their initial hospitalization with a gastrostomy tube following a Norwood procedure. However, the expectation of feeding dysfunction may bias the clinician to ignore other causes of feed intolerance. Here, we present three cases of infantile hypertrophic pyloric stenosis (IHPS) to

illustrate that while feeding dysfunction may be expected in children with cyanotic heart disease, other structural causes should be evaluated.

CASE REPORT

Patient 1 is an 8-month-old male with HLHS who received prostaglandin until his Norwood procedure at 2-weeks of age. Postoperatively, he remained dependent on nasogastric feeding due to concerns of aspiration, which led to consideration for a gastrostomy tube. As part of the preoperative workup, an upper gastrointestinal (GI) imaging series showed esophageal dysmotility but normal anatomy of the stomach and duodenum, and the infant underwent a laparoscopic gastrostomy tube placement at the age of 5 weeks. Over the next 3 weeks, he developed progressively worsening forceful emesis. Fluoroscopic evaluation, which was performed to evaluate the gastrostomy tube and the pylorus, showed normal filling of the stomach with delayed passage of contrast out of the stomach. An abdominal ultrasound ultimately confirmed the diagnosis of IHPS. Under general anesthesia, the patient underwent pyloromyotomy. Intraoperatively, the pylorus measured 25 mm in length and was clearly hypertrophic. Postoperative course was uneventful, and he was discharged home 6 days following pyloromyotomy. At follow-up, he showed excessive unanticipated growth and his feeding regimen was decreased accordingly.

Patient 2 is a 6-week-old male with HLHS and intact septum. He underwent placement of an atrial stent shortly after birth. This patient was maintained on prostaglandin for 13 days until atrial septectomy and subsequent placement of a patent ductus arteriosus stent and bilateral pulmonary artery bands (“hybrid procedure”). His postoperative course was complicated by necrotizing enterocolitis. After completing antibiotics and reinitiating feeds, he had persistent feeding difficulty and was diagnosed with severe pathologic reflux requiring nasojejunal feeds. At 1-month of age, fluoroscopic upper GI series imaging and an ultrasound of the pylorus showed no evidence of IHPS but did show markedly delayed gastric emptying. The patient was started on a promotility agent, erythromycin. Because of persistent emesis, metoclopramide was added, and a repeat ultrasound was obtained 1 week later. The new study demonstrated a hypertrophic pylorus with a wall muscle thickness of 4 mm and a channel length of 2 cm. A pyloromyotomy was performed under general anesthesia. The patient recovered well from the procedure and remains in the hospital recovering from acute hypoxemic respiratory failure and pulmonary hypertension. His reflux resolved, but he requires nasogastric feedings and speech therapy because of poor oro-motor skills.

Patient 3 is a 5-week-old boy with double-outlet right ventricle with subpulmonic ventricular septal defect, L-TGA, and interrupted aortic arch (IAA), type A. His left-sided right ventricle showed mild to moderate hypoplasia. He was maintained on prostaglandin for 21 days. Following repair of his IAA and placement of pulmonary artery bands, he was placed on nasogastric feeds because of concern of aspiration. Following discharge home, he developed worsening episodes of nonbilious vomiting. He was readmitted, and ultrasound showed a 5 mm thick pylorus with a length of 2.1 cm consistent with IHPS. He did well postoperatively with resolution of vomiting and has since undergone biventricular repair.

DISCUSSION

As we have noted, feeding complications are documented at a higher incidence particularly in infants with cyanotic heart disease. Boctor *et al*. determined that weight gain in infants with congenital cardiac defects is suboptimal following cardiac surgery, with a median overall daily weight loss of 11 g/day in a retrospective review of 24 infants, including two with HLHS.[6] Many centers have developed feeding algorithms for children with complex congenital heart disease, specifically complex left ventricular outflow tract obstructions such as HLHS status post-Norwood. It is perhaps not surprising considering that some degree of feeding dysfunction is expected in this subset of patients that other possible etiologies of feeding dysfunction may go unnoticed.

We report three cases of feeding dysfunction in children with single ventricle physiology secondary to underlying IHPS. The thickened pylorus leads to gastric outlet obstruction, which, in turn, leads to emesis and loss of electrolytes. Children with IHPS typically present between 4 and 6 weeks of age with projectile, nonbilious vomiting of feeds. They may show visible abdominal peristaltic waves against the fixed obstruction. IHPS may be confirmed with ultrasonography or contrast-swallow upper GI imaging series. Typical diagnosis criteria include a pyloric thickness >3 mm (100% sensitivity, 99% specificity) and a pyloric length >15 mm (100% sensitivity, 97% specificity).[7] The etiology of IHPS is largely unknown.[8] There is evidence for involvement of at least two loci on chromosomes 3 and 5 based on interfamilial association analysis studies.[9] IHPS may also occur as part of an overall syndrome such as Smith–Lemli–Opitz.

A pyloric stenosis-like entity, prostaglandin-induced foveolar hyperplasia, has been described in infants who are on prolonged prostaglandins to maintain ductal patency for cyanotic heart disease.[10] This interesting phenomenon can coexist with IHPS and is diagnosed through similar testing, which reveals pronounced gastric lobulations and progressive elongation of the antropyloric channel with or without wall thickening. It creates a similar gastric outlet obstruction and clinical picture. In some cases, discontinuation of prostaglandin alone is adequate to treat prostaglandin-induced foveolar hyperplasia.[11] When foveolar hyperplasia exists in conjunction with thickening of the pylorus, surgery is the primary treatment option. Symptoms in the children described here persisted despite first stage cardiac surgery and discontinuation of prostaglandin. Because the pyloric thickness measurements of these patients were consistent with IHPS and because of symptomatic improvement following pyloromyotomy, additional evaluation, such as histopathology of the gastric foveolar cells, was not felt to be indicated.

In this report, we have described three patients with left-sided obstructive lesions who developed IHPS. While commonalities exist between these patients, we cannot identify any causality with such small numbers. It is important to note that although feeding dysfunction is expected in this patient population, other structural etiologies such as pyloric stenosis can coexist. We recommend that pyloric stenosis remains in the differential for feed intolerance postcardiac surgery and for appropriate diagnostic testing to be carried out to diagnose this entity.

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Conflicts of interest

There are no conflicts of interest.

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