

## Neonatal Appendicitis Presenting as Bilioid Emesis and Septic Shock

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### INTRODUCTION

The incidence of neonatal intestinal obstruction is estimated to be approximately 1 in 2,000 live births, often presenting with symptoms of bilioid emesis.<sup>1</sup> A surgical indication can be found in up to 38% of infants when presenting with bilioid emesis.<sup>2</sup> Plain radiography, contrast radiologic studies, and ultrasonography are imaging studies commonly used in the evaluation of patients presenting with bilioid emesis, with upper gastrointestinal series being the preferred modality.<sup>3</sup> The causes of bilioid emesis in a neonate are broad and include, but are not limited to, Hirschsprung's disease, bowel atresia, malrotation, and volvulus; the differential rarely includes appendicitis.

Neonatal appendicitis is a rare condition with a reported incidence rate of 0.04 - 0.2%.<sup>4</sup> Classical findings of appendicitis, such as decreased appetite, fever, abdominal pain, and leukocytosis, are often absent, resulting in a delay in diagnosis and management.<sup>4,5</sup> This nebulous presentation rarely results in a preoperative diagnosis and often is discovered on exploratory laparotomy or postmortem examination.<sup>4</sup> Accurate mortality rates are difficult to establish, however, they have improved. Rates in the early 2000s were 23 - 28%, while one retrospective review of 31 cases from 2001 to 2018 showed 0% mortality rate.<sup>6-8</sup> Perforation plays a significant role in predicting the prognosis of neonatal patients with appendicitis.<sup>4,5</sup> Because of the underdeveloped omentum found in neonates, it is thought that any form of purulent material cannot be processed and leads to diffuse peritonitis.<sup>9</sup> Clinical deterioration often prompts surgical intervention leading to the diagnosis. We present a unique case of a neonate with bilioid emesis caused by appendicitis that ultimately lead to multi-organ system failure.

### CASE REPORT

A 38-week gestational age female with a perinatal history complicated by polyhydramnios was admitted to the special care nursery due to hypoglycemia. After initial requirement for dextrose containing fluids, she was able to maintain euglycemia by infant formula at four days of life (DOL). She was discharged home without any signs of feeding intolerance or bowel irregularity. At DOL five, she was brought to the emergency department by the parents with concerns for fussiness and poor feeding. An abdominal radiograph was unremarkable, and her formula was changed, and she was able to tolerate 33 ml by mouth with resolution of perceived fussiness. Her vital signs were temperature of 99.8°F, heart rate of 129 bpm, respiratory rate of 32 breaths/min, and

an oxygen saturation of 100% in ambient air. She was discharged home with anticipatory guidance the same night.

On DOL eight, she was brought back to the emergency department with continued fussiness, poor oral intake, and bilioid emesis. An abdominal ultrasound showed small to moderate ascites, but no other abnormalities. An upper gastrointestinal radiograph under fluoroscopy showed no evidence of malrotation or volvulus. However, it revealed episodes of severe reflux to the upper esophagus and multiple dilated loops of small bowel concerning for distal bowel obstruction. The decision was made for emergent exploratory laparotomy after surgical consultation. Intra-operative findings included a contaminated peritoneal cavity with purulent drainage, turbid ascites, and inflammatory small bowel obstruction of the ileum in the right lower quadrant with a perforated necrotic appendix. Appendectomy was performed, peritoneal cultures were obtained, and the abdomen was closed.

She was admitted post-operatively to the Pediatric Intensive Care Unit (PICU) for monitoring. Her initial vital signs at admission were temperature of 97.5°F, heart rate of 172 bpm, respiratory rate of 65 breaths/min, blood pressure of 89/66 mmHg, and an oxygen saturation of 96%. Admission labs are shown in Table 1. Physical exam revealed an ill appearing infant with subcostal retractions and grunting, decreased peripheral pulses with a capillary refill of three seconds, absent bowel sounds, a distended abdomen that was firm to palpation in the lower quadrant, and was responsive to noxious stimuli with moaning.

**Table 1. Labs obtained prior to initial exploratory laparotomy.**

Sodium (mmol/L)	136	WBC (k/cumm)	22.6
Potassium (mmol/L)	6.9	Hemoglobin (g/dL)	19.8
Chloride (mmol/L)	102	Hematocrit (%)	55.5
Anion Gap (mmol/L)	22	Platelet (k/cumm)	323
BUN (mg/dL)	29	Creatinine (mg/dL)	0.70
Glucose (mg/dL)	63	Calcium (mg/dL)	6.4
AST (units/L)	117	ALT (units/L)	160
Albumin (g/L)	2.4		

Fluid resuscitation up to 80 mL/kg in aliquots with crystalloids resulted in minimal improvement in perfusion and lactic acid. She soon required tracheal intubation and invasive mechanical ventilation for optimizing oxygen delivery. She was initiated on vasoactive agents norepinephrine and epinephrine for fluid refractory shock and received stress dose hydrocortisone intravenously. The surgical team was reconsulted given her persistent lactatemia, metabolic acidosis, and increasing doses of vasoactive infusions. Her abdomen remained distended, taut, and discolored, and she remained anuric during this time. Given concerns for intra-abdominal hypertension and abdominal compartment syndrome, a repeat exploratory laparotomy was performed 18 hours post-admission and fascial planes were left open with only skin closure for abdominal decompression.

Table 2 shows the progression of labs from before and after second exploratory laparotomy. Eventually, the decision was made to transfer her to a quaternary care center for initiation of veno-arterial extracorporeal membrane oxygenation (ECMO) and continuous renal replacement therapy for multi-organ failure refractory to conventional therapy. Transfer occurred at around 19 hours postoperatively. The

patient unfortunately had cardiac arrest upon arrival to the outside hospital. Return of spontaneous circulation was achieved after several minutes of resuscitation and decompression of the abdomen with placement of intestinal loops in silo. The bowel appeared ischemic and dusky and the decision was made to not pursue ECMO given poor prognosis. Parents chose discontinuation of life sustaining measures and patient eventually died.

**Table 2. Lab trends comparing pre- and post- second exploratory laparotomy.\***

	Pre-Op Labs			Post-Op Labs	
	At 10 hours	At 16 hours	At 18 hours	At 21 hours	At 24 hours
Lactate (mmol/L)	7.1	5.0	7.2	9.8	12.1
Sodium (mmol/L)	144	148		151	153
Potassium (mmol/L)	5.1	5.6		5.3	4.4
Chloride (mmol/L)	109	116		116	116
CO2 (mmol/L)	14	13			
Anion Gap (mmol/L)	21	19		23	22
BUN (mg/dL)	28	31		30	32
Creatinine (mg/dL)	1.16	0.71		1.06	1.10
Calcium (mg/dL)	5.4	<5.0		7.1	7.0
Calcium (ionized) (mg/dL)		3.3		4.1	3.9
Magnesium (mg/dL)		2.5			
AST (units/L)				764	611
ALT (units/L)				231	171
Albumin (g/dL)	1.3		0.7	1.4	2.1
Venous blood gas					
pH	7.08	7.04	6.91	7.09	7.12
pCO2 (mmHg)	47	42	40	34	41
HCO3 (mEq/L)	13.6	11.0	7.8	9.9	13
Base Excess (mEq/L)	-16.3	-19.2	-24.9	-19	-15.7
Prothrombin (s)		32.7			
INR		2.8			
aPTT (s)		104			
Fibrinogen (mg/dL)		133			

\*Time indicated is in hours from admission. Labs closest to the indicated hour are presented. Her initial laparotomy occurred at seven hours post admission, with her repeat surgery occurring 18 hours post admission.

## DISCUSSION

Neonatal appendicitis is a rare but potentially life-threatening condition. This report discussed the unique presentation of a patient that experienced rapid deterioration following admission to the pediatric intensive care unit. Despite the patient's clinical course, this case illustrated the importance of early recognition of neonatal appendicitis and some of the challenges associated with the management of severe shock in the neonatal population.

While appendicitis is seen commonly in children, it rarely causes bilious emesis and rarely affects neonates. This is thought to be due to the wide opening of the appendix in the newborn, infrequent abdominal or respiratory infections, a liquid diet, and recumbent positioning.<sup>10</sup> The wide opening prevents obstruction with fecaliths or other food

particles. With lower rates of abdominal and respiratory infections, there is less hyperplasia of lymphoid tissue in the gut, which potentially serves as a protective factor from obstruction of the appendix. Unfortunately, the neonatal appendix tissue is thin-walled and fragile, making it prone to perforation.<sup>6</sup> The neonatal immune system also is inexperienced and responds poorly to overwhelming infections.<sup>11</sup> The omentum is underdeveloped and is unable to absorb purulent material, resulting in widespread dissemination after perforation of the infection to the small neonatal abdominal cavity.<sup>9</sup> Also contributing is the difficulty in making an early diagnosis in this patient population. Most diagnoses occur postoperatively after signs of peritonitis, abdominal distension, or bilious emesis appear.<sup>7,12,13</sup>

Our case followed a similar pattern, whereby an acutely perforated appendix was discovered upon exploratory laparotomy to investigate the source of obstruction. Imaging modalities in neonates provide little assistance in identifying appendicitis, but are helpful in detecting complications, such as pneumoperitoneum on x-ray, free abdominal fluid on ultrasonography, or upper gastrointestinal x-rays with contrast showing bowel obstruction.<sup>11</sup> Signs suggestive of bowel obstruction were identified in our case, which prompted further surgical intervention.

Multiple hypotheses have been made towards the etiology of neonatal appendicitis. One theory is from overdistension of colonic tissue and appendiceal tissue secondary to Hirschsprung disease.<sup>13</sup> There was no evidence of colonic distension in our patient and she lacked the classical findings of delayed meconium passage and had multiple bowel movements in the first day of life. Another suggested theory is a localized necrotizing enterocolitis secondary to necrosis of the appendiceal wall.<sup>14</sup> Even though this is plausible, the patient in our case was not premature, which is a major predisposition to the condition.

Although difficult to assess the presentation of neonatal appendicitis, some clinical features may be indicative of the condition. Early symptoms, as also seen in our case, include irritability and poor feeding while late findings include abdominal distension, abdominal guarding, and bilious emesis.<sup>4-6,15,16</sup> Clinicians should have a high index of suspicion for early diagnosis, management, and prevent sequelae of complications.

## CONCLUSIONS

This case illustrated the clinical presentation and course of a term neonate with acute appendicitis. Diagnosis is made by prompt surgical evaluation in addition to clinical and radiographic findings. Complications can include sepsis, abdominal compartment syndrome, multiple organ dysfunction syndrome, and death in rare cases. Albeit a rare cause of bilious emesis, neonatal appendicitis is an important differential for bilious emesis in the neonatal period.

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