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# Post Necrotizing Enterocolitis Intestinal Stricture- A mimicker of Hirschsprung'S disease

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**Abstract**

Necrotizing enterocolitis (NEC) is an inflammatory disease process of the gastrointestinal tract commonly affecting premature (< 37 weeks) infants. The NEC mortality rate has decreased from 50% to currently up to 20 % because of a better available understanding of disease and management. The sequel of post-NEC complications viz strictures can be isolated or multiple. These strictures can be managed via excision of the stricture bowel and primary end-to-end enteric anastomosis. The other approach is staged surgical repair: excision of the strictured bowel segment with proximal diversion enterostomy, delayed water-soluble contrast distal-loopogram with or without contrast enema, followed by delayed closure of stoma. The author discussed a case of Post-NEC stricture relevant review of the literature.

**Keywords**: Necrotizing enterocolitis, post-NEC complications, post-NEC stricture Predictive factors for stricture, Recurrent post-NEC strictures, recurrent NEC

## Introduction

Necrotizing enterocolitis (NEC) is an inflammatory disease process of the gastrointestinal tract. NEC is commonly seen in neonatal intensive care units affecting premature (< 37 weeks) infants (1). The mainstay of management in NEC is medical intervention, consisting of abdominal decompression by nasogastric tube, bowel rest by keeping nil per orally, broad-spectrum intravenous antibiotics, and total parenteral nutrition (in cases prolonged bowel rest is needed for more than 14 days). Emergency surgery is required in infants with parietal wall erythema, intestinal perforation, portal vein gas, fixed bowel loops on abdominal X-ray, severe NEC, and failure of medical management (2)(3). Surgical management includes exploratory laparotomy, enterostomy, resection anastomosis, and 'clip and drop' techniques or bedside peritoneal drainage

Advanced medical management of NEC decreases the mortality of neonates from 50% to 20 %(4) (5). The mortality of neonates who underwent any surgical procedure varies from 31 % to 67% in different series(2) (4).

The patients having sequelae of post-NEC complications viz intestinal strictures increased from 5% to 35% (6). These intestinal strictures might be isolated or at multiple sites. Isolated Post-NEC stricture occurs in 75% of cases(7) (8). The multiple Post-NEC strictures might present simultaneously or at some time interval. The author critically appraised a case of recurrent post-NEC stricture with a discussion of the existing literature.

## Case Discussion:

**Case 1:** A preterm (36 weeks Gestational age), normal vaginal delivered, female, having birth weight 2.2 Kg, presented with abdominal distension and

bilious vomiting on the 14th day of life. The baby was managed at a peripheral hospital. The diagnosis of late-onset Modified Bell's stage II-B NEC was made based on clinic-laboratory parameters (figure 1) Table 1. Hospital Record analysis of the baby revealed medical management included, nil per oral, intravenous fluid, and intravenous antibiotics covering Gram-positive, Gram-negative, and anaerobe coverage. Total parental nutrition, inotropes, and/or mechanical ventilation were not required during the first index admission. Blood investigations showed the baby was sickle-cell trait positive. The baby improved on conservative management and was discharged on the 30th day of life.

On the 40th day of life, the baby presented to our hospital with clinical features of distal intestinal obstruction. The X-ray abdomen showed multiple air fluid levels. The baby had grade IV of Indian Association of paediatric classification (IAP) malnutrition, with hypoalbuminemia, and electrolyte imbalance (hyperkalemia). The baby failed to improve on conservative measures. Water-soluble Contrast enema showed narrowing at the sigmoid colon, with reversal of rectosigmoid ratio, dilated descending colon, and delayed retention of contrast. These findings raised suspicion of Hirschsprung's disease (figure 2). The baby failed to improve on rectal washes. The anorectal manometry facility was not available, so the decision for exploratory laparotomy was made. Laparotomy was performed through the left lower quadrant hockey stick incision. under general anesthesia. A probe patent stricture of 1 cm was identified at the recto-sigmoid junction. The strictured sigmoid colon was excised and a double-barrel sigmoid colostomy was made.

The histology of excised sigmoid stricture showed adequate ganglionic cells, with nonspecific inflammatory infiltrates.

Postoperatively 15 days were uneventful, but after that patient again developed features of intestinal obstruction. Histopathology slides were reviewed in whole circumference, but ganglion cell was present in adequate number, without any features of nerve bundle hyperplasia. The baby was not responding to the colonic washes and bowel rest.

The baby underwent a second exploratory laparotomy on the 65th day of life. The author found new stricture at ascending colon just distal to the ileocecal junction. Intraoperatively baby was hemodynamically unstable, so, a diverting loop ileostomy was made (figure 3).

After 2nd surgery baby developed complete wound dehiscence, needing abdominal closure under general anesthesia. Thereafter baby recovered from sepsis, tolerating orally well and gaining weight.

The baby was discharged on the 75th day of life with loop ileostomy and sigmoid double barrel colostomy in situ. In follow-up of the outdoor department, the baby developed prolapsed loop ileostomy. The baby was gradually put on a high-protein diet with vitamin supplementation.

At 3 months of age, the baby gained a weight of 4 Kg. The retrograde colostogram did through the proximal stoma of the sigmoid colostomy, which demonstrated the persistence of stricture at ascending colon just distal to the caecum.

The fourth laparotomy was done under general anesthesia. The strictured ascending colon, cecum, and ileocecal junction were excised with ileo-transverse anastomosis and closure of sigmoid colostomy.

Histology of excised ascending colonic stricture showed nonspecific inflammation with adequate ganglion cells without any nerve hypertrophy. There was no feature of Cytomegalovirus (CMV) inclusions body in the strictured ascending colon.

**Results:** The baby is healthy, and thriving well within 3 yrs of follow-up. There was no adhesive intestinal obstruction or further evidence of strictures in the entire bowel. The neurodevelopmental and sensory development of the child is normal to date.

**Discussion:**

The cause of mortality in acute NEC is extensive bowel involvement, NEC-related sepsis, and multiple organ failure syndrome(9)(10). The delayed causes of mortality in survivors of acute NEC are late Intestinal failure or intestinal failure-associated liver diseases.

Late intestinal failure is a chronic condition characterized by a reduction in functional intestinal mass required for adequate digestion and absorption of nutrients, fluids, and growth requirements. Intestinal failure might be due to short bowel



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syndrome after surgery or other congenital conditions that reduce the mucosal surface area.

Delayed intestinal failure-associated Liver disease usually develops after receiving parenteral nutrition for more than 100 days. This liver failure occurs in 85% of NEC babies receiving TPN.(4)

Post-NEC strictures are delayed consequences of NEC. An extensive online search on Medline with keywords post-NEC stricture, Predictive factors, recurrent strictures, and recurrent NEC helped in the Critical appraisal of the case as follows.

Post-NEC strictures occur due to the healing process that follows ischemic injury to the inner muscular layer of the intestine(11). The stricture is suspected if the baby develops food intolerance, recurrent abdominal distension, and high gastric aspirates.

The most common site for post-NEC stricture is the colon and distal ileum (75%)(9). Post-NEC strictures manifest in the $5^{th}$-$7^{th}$ weeks or up to the $13^{th}$ weeks after the acute episode of NEC(9). The stricture develops irrespective of primary management strategies (medical and/ or surgical) of an acute episode of NEC(12)(13).

The radiological features of intestinal obstruction on water-soluble contrast enema confirm the diagnosis and location of post-NEC stricture.

If the Contrast enema is normal, an upper gastrointestinal study with small bowel follow-through is recommended to identify the stricture(14). In our case contrast, the enema was mimicking with a transition zone of hirschsprung's disease. On exploratory laparotomy, the author found the rectosigmoid stricture with normal ganglion cells in the excised specimen.

Risk factors of stricture formation are difficult to predict following acute episodes of NEC. However, late-onset NEC (>14 days), increased plateletcrit, leucocytosis, and C-Reactive Protein might be associated with an increased risk of NEC strictures (9)(10). Except for leucocytosis and late-onset NEC, our case didn't have another risk factor for the development of post-NEC stricture.

The most common etiology of Post-NEC strictures is chronic progressive inflammation at multiple sites in the bowel(9)(15). The surgeons must be aware that intestinal strictures might also be sequelae of inadequate resection of the necrotic bowel during primary surgical management in babies suffering from acute NEC (9)(15).

The management of the Post-NEC stricture, in the 1970s and 1980s, was preferably stoma formation in the initial step, alongside primary anastomosis(9). Post- NEC strictures were excised with proximal diversion ileostomy/colostomy, given the possible development of new stricture formation. The etiology of new stricture formation was a persistent inflammatory process in the bowel maximum up to 3 months of age in children(15). Our case might develop a second stricture because of the ongoing inflammatory process. Further, the proponents of surgeons favoring stoma formation justify themselves on the fact that, patients developing post-NEC stricture are in sepsis and malnutrition, so primary anastomosis is warranted. Primary anastomosis is also avoided in circumstances of the gross discrepancy between bowel ends.

Contrarily few case series showed primary bowel anastomosis in Post-NEC stricture, which gave good and promising results with a decrease in morbidity related to stoma and bowel disparity (15). There is a very low risk of same-site recurrent intestinal strictures or another-site new Post-NEC intestinal strictures.

To the best of the author's search literature role of stricturoplasty in post-NEC stricture has not been described to date.

Other long-term complications of NEC are deafness, developmental delay, multiple food allergies, and recurrent gastrointestinal challenges from short gut syndrome is under evaluation by the child in close follow-up (16).

**Conclusions**

Adequate excision of Post-NEC strictured bowel segments with a stoma can be a safe option.

The contrast loopogram (colostogram/ileostogram) is recommended before stoma closure, because NEC is a progressive inflammation of the entire gastrointestinal tract, and the possibility of new stricture or missed stricture must always be ruled out.

Further, given the rarity of the entity well planned multicenter Randomized Control Trial or/and a prospective study comparing different surgical



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approaches in cases of Post-NEC stricture is needed to confirm the impact of a specific strategy.

**Take away Message**

- Adequate excision of Post NEC stricture segment with primary anastomosis of the bowel can be a safe option. However, eagle eyes should be kept on the possibility of the appearance of new stricture.
- Even in cases of diverting soma distal colostogram is a must before stoma reversal to rule out undiagnosed stricture that may appear after diversion surgery of primary stricture

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Table legends:

Table 1: Baseline characteristic and Hematological investigations of the patient during first index admission and second admission

| **Baseline characteristic of the patient** | first index admission (day 14th of life) | Second admission (day ) |
|---|---|---|
| Gestational age in weeks | 36 week | - |
| Gender | female | - |
| Birth weight | 2.2 kg | - |
| APGAR Score at 5 minutes of Birth | 8 | - |
| Age at diagnosis of NEC | 14th day of birth | - |
| Maternal age | 28 yrs | - |
| Abdominal distension | Present | Present |
| letharginess | Present | Present |
| Bilious vomiting | Present | absent |
| Blood in stool | microscopic | absent |
| Absent bowel sound | absent | absent |
| Abdominal tenderness | Present | absent |
| Generalized peritonitis | absent | absent |
| Abdominal wall cellulitis | absent | absent |
| Abdominal mass | absent | absent |
| Hypotension | absent | absent |
| DIC | absent | absent |
| Assisted Ventilation | Not required | Not required |
| Comorbid conditions | Sickle Cell trait | Sickle Cell trait |
| Other Congenital anomalies | absent | - |
| **Hematological investigations** | | |
| Total white cell count (onset of first symptoms) $10^9$/L | 11.8 | 13.8 |
| Thrombocytopenia (onset of first symptoms) $10^9$/L | 280.2 | 220.0 |
| C- reactive protein (onset of first symptoms) mg/L(normal = 1-10) | 18 | 24 |
| S Procalcitonin (ng/ml) (normal = 0-0.5 ) | 4 | 6 |
| S Albumin (N= 30-50 g/L) | 24 | 20 |
| Metabolic acidosis (onset of first symptoms) | absent | absent |
| Serum potassium (onset of first symptoms) mEq/L (normal =5.0-6 ) | 4.5 | 5.5 |
| Serum Sodium (onset of first symptoms) mEq/L (normal =139-162 ) | 152 | 155 |
| Serum calcium (onset of first symptoms) mmol/L (normal = 1.5- | 1.1 | 1.3 |



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| | | |
|---|---|---|
| 2.4) | | |
| Lactic acidosis (onset of first symptoms) | absent | absent |
| Serum bicarbonate(onset of first symptoms) mEq/L (normal 22-26) | 28 | 21 |
| Positive Blood Culture (onset of first symptoms) | Positive | positive |
| Blood transfusion (Packed Red Blood Cell) | Required | Required |
| Surgical management | Not required | Operated for stricture |

Figure legends:

**Figure Legends**

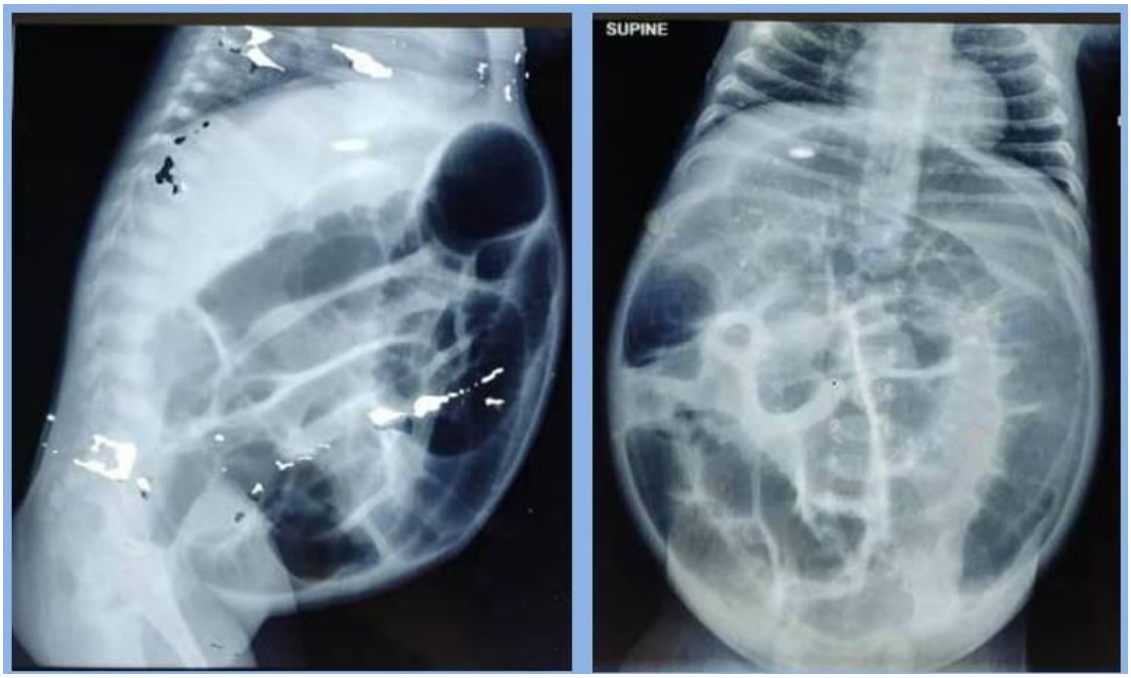


Figure 1: X-ray abdomen (supine): (a) lateral view, (b) anteroposterior view showing dilate bowel loops distal intestinal obstruction



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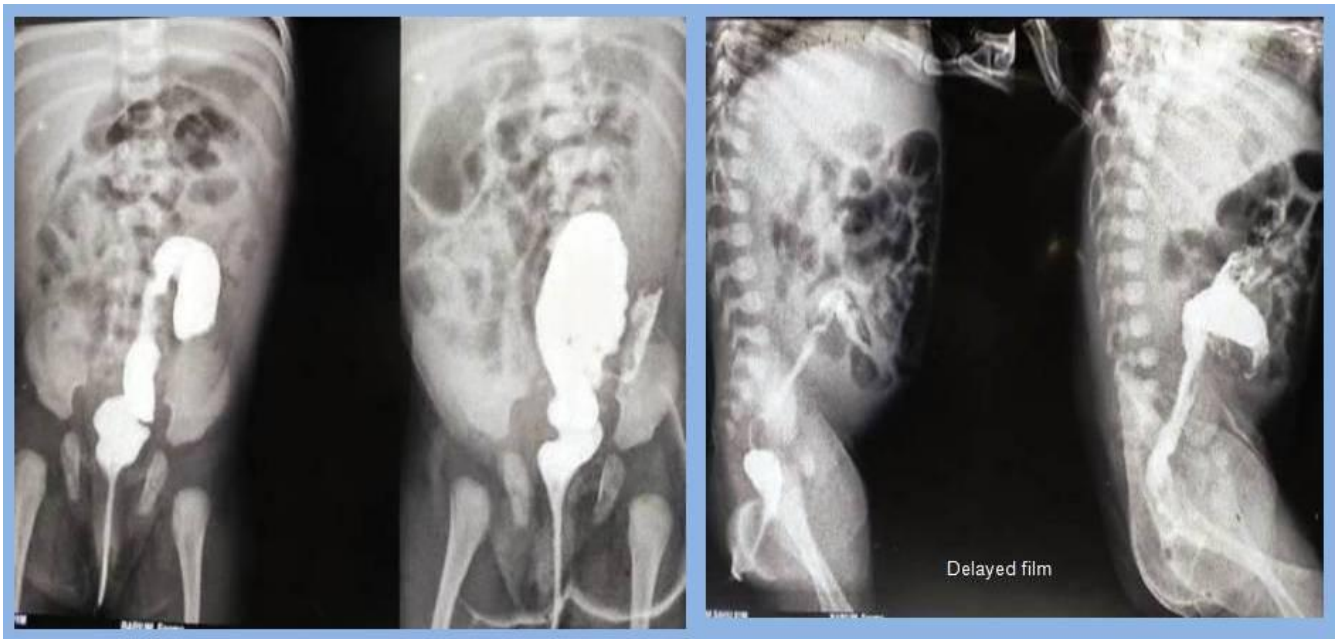


Figure 2: Water-soluble contrast enema: (a) showing narrowing at sigmoid colon, dilated proximal colon, and narrow rectum (b) Delayed films (12 hours) of Contrast enema showed retention of contrast in the rectum and sigmoid

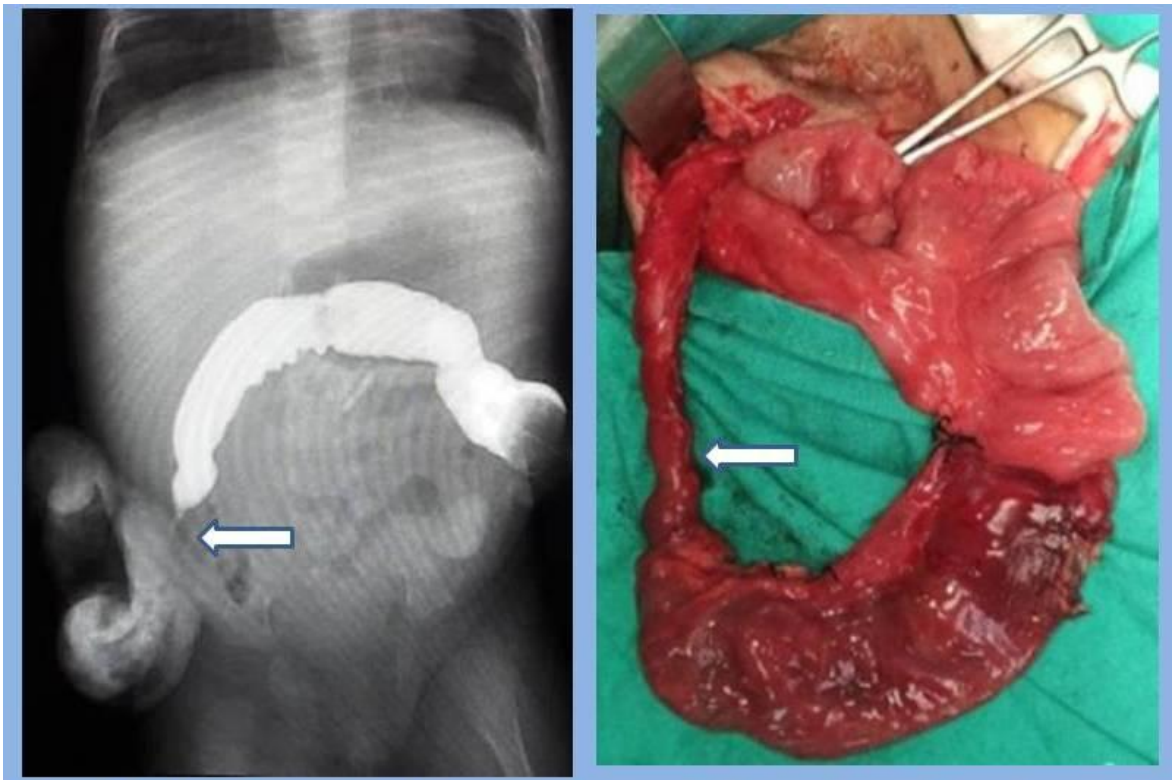

Figure 3: (A) Retrograde colonogram through sigmoid colostomy showing stricture at ascending colon (White arrow), with soft tissue shadow of prolapsing ileostomy (B) Probe Patent, 2 cm Stricture at ascending colon (white arrow)



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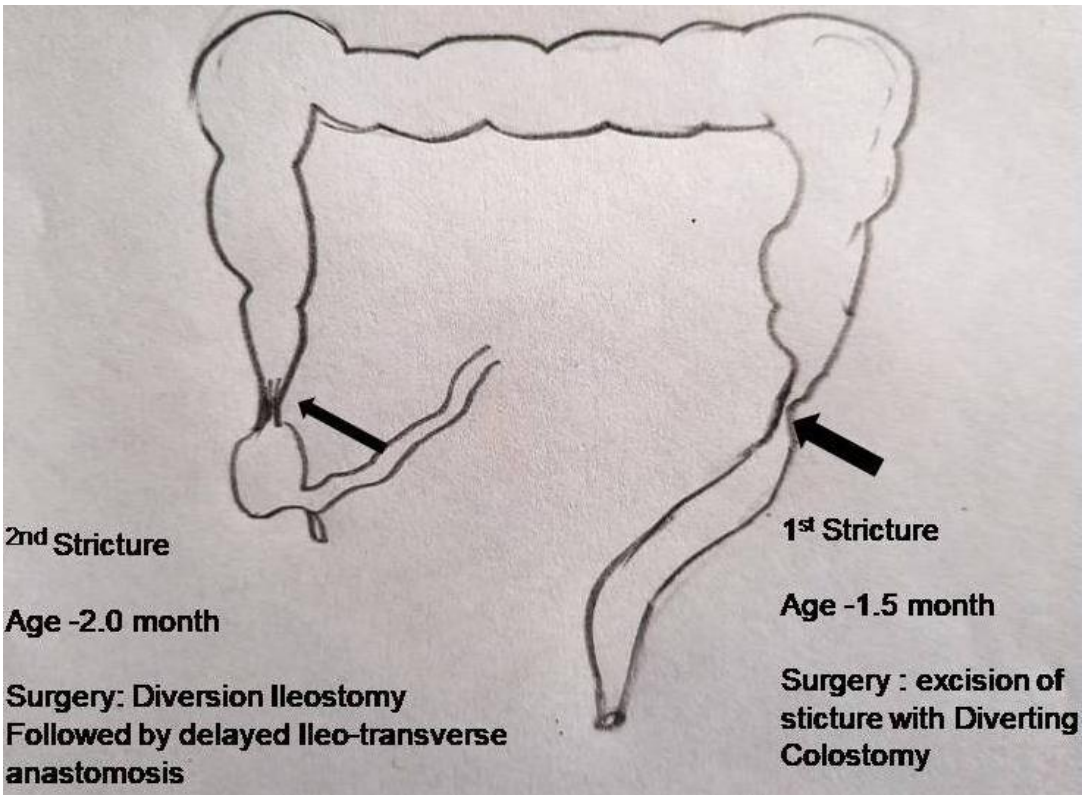


Figure 4: Schematic presentation of staged management of multiple progressive post-NEC strictures in the colon

- Disclosure - none to declare
- Conflict of interest - none
- Previous presentation - The case report has been presented as Scientific Paper at the 46th National conference of the Indian Association of Pediatric Surgeons, India 2020
- Submitted as a manuscript or accepted in any journal - No
- Acknowledgement: none.



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