Case Series

Congenital Neonatal Colonic Atresias Arising in Watershed Areas of the Colonic Blood Supply

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There are certain junctional weak points in the colonic marginal artery blood supply, known as watershed areas, which result from congenital incomplete development of anastomoses of the marginal arteries. These critical points in the marginal arcade are more vulnerable to consequences of ischemic injury than other parts of the marginal arterial arcade. We report herein the series of five patients who developed colonic atresia well localized in the rectosigmoid region at Sudeck's point in two patients, in the splenic flexure region at Griffith's point in one patient and in the cecum-ascending colon junction in one patient respectively and a very interesting case of wide spread multiple junctions at middle colic and inferior mesenteric artery distribution type 3 atresia in one case. This report and our review of the literature suggest that watershed areas, including Sudeck's point or rectosigmoid junction area, the splenic flexure or Griffith's point and the ileocecal region between ileal and colic branches and combinations thereof, are high-risk regions for the development of colonic atresia and rarely the vascular insult can be major and can involve key branch of marginal arcade leading to short colon syndrome. We have treated all different approaches with one stage, two stage and three stage open, laparoscopic and minimal invasive periumbilical approach. All three approaches and single stage or staged approaches are safe and effective in colonic atresias depending on the case and available resources. An attempt to save ileocecal valve, appendix and cecum with ceco-coloplasty in the initial stage did not work and had to finally perform subtotal resection of the dilated cecum and part of the ascending colon with preservation of the ileocecal valve and the appendix at corrective surgery in the right colonic atresia case. We had motility disorders associated with two of our cases.

[*J Indian Med Assoc* 2024; **122(2):** 55-60]

Key words **: Colonic Atresia, Griffith Point, Sudeck Point, Middle Colic Artery, Neonatal Intestinal Obstruction, Left Transverse Colostomy, Laparoscopy.**

Isolated Colonic Atresia in a neonate is exceedingly rare, least common intestinal atresia, poses several diagnostic and therapeutic challenges for successful and safe outcome of ideal treatment which has consequences of life long implications[1-11]. The Colonic Atresias are different from other intestinal atresias as there is a huge disproportion between the proximal and distal segments and hence, more likely to be managed by staged approach rather than single stage repair. Plain radiograph and contrast enema should help establish the diagnosis. Here, we present a series of neonatal colonic atresia cases with its all-associated risks, with a review of the literature focusing on preservation of ileocecal valve, appendix and colon with emphasis on innovative minimal

Editor's Comment :

- The vascular theory of colonic atresia is well supported by occurrence at three critical junctional weak points called watershed areas of marginal colonic artery anastomosis as congenital anomaly between different branches is a new finding and objective evidence.
- Association of Hirschsprung's disease with colonic atresia is well known but we have reported motility disorders as variant Hirschsprung's disease.
- The watershed areas, including Sudeck's point or rectosigmoid junction area, the splenic flexure or Griffith's point, and the ileocecal region between ileal and colic branches and combinations thereof, are high-risk regions for the development of colonic atresia.
- All three approaches (open, periumbilical and laparoscopic) and single stage or staged approaches are safe and effective in colonic atresias depending on the case and available resources.

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Received on : 10/02/2022
Accepted on : 20/02/2022

invasive surgery and its peri-operative care. We aim to contribute to the awareness of the existence of these rare lesions and safe and successful surgical management.

Case 1

A 2080 grams baby girl was delivered normally at 36 weeks. Two days after birth she developed abdominal distension, failure to pass meconium and bilious vomiting. Physical examination and abdominal X-ray showed bowel obstruction with no gas in the rectum (Fig 1A). Contrast enema confirmed Colonic Atresia at rectosigmoid junction (Fig 1B).

Left transverse colostomy with biopsy at colostomy site and rectal suction biopsies, to rule out associated Hirschsprung's disease, were performed in the neonatal period uneventfully. The histological examination of both biopsies showed immature ganglion cells. At definitive surgery, type 2 rectosigmoid Colonic Atresia was resected and end to end Anastomosis was performed at 4 months after repeat rectal suction biopsy showed normal mature ganglion cells. Postoperative colostogram was normal and Colostomy closure was done at 6 months of age uneventfully. Follow up was uneventful and the patient is now 16 years old with normal feeding and transit.

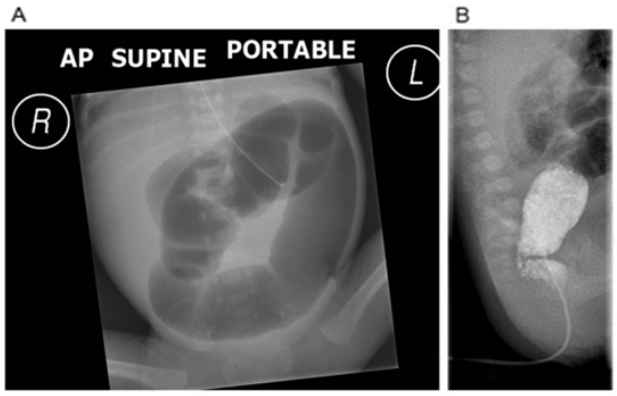


Fig 1 — Plain abdominal radiograph and contrast enema of Case 1

Case 2

A full-term male neonate weighing 3450 grams was delivered by normal vaginal delivery. On the first day of life, he was admitted with bilious vomiting and mild abdominal fullness.Chest radiograph was normal. Plain radiograph showed dilated small bowel and no gas was visualized within the rectum suggesting a distal bowel obstruction (Fig 2A). Bowel atresia, a meconium ileus or long segment Hirschsprung's disease was a probability. Water soluble contrast enema passed easily up to a point probably involving the splenic flexure but without any significant meconium or bowel content present within the lumen of the distal microcolon. It would not pass any further than this and no evidence of caliber change anywhere to suggest Hirschsprung's disease. Appearances would suggest a Colonic Atresia with

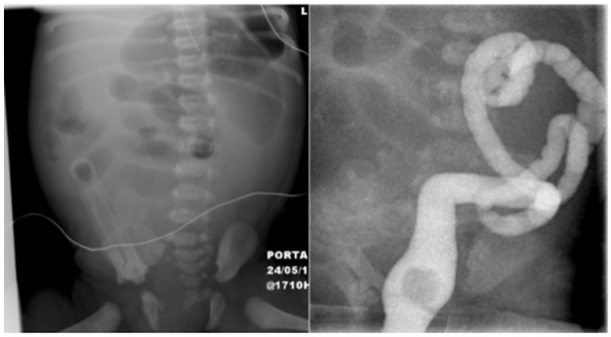


Fig 2 — Plain abdominal radiograph and contrast enema of Case 2

adjacent proximal bowel loops did appear dilated (Fig 2B).

Rectal suction biopsy showed few ganglion cells but no hypertrophy of nerve fibers and normal acetylcholinesterase suggestive of Hypoganglionosis. As there was no significant abdominal distention and good weight term baby, an option for laparoscopic evaluation and exteriorization of the atresia at one of the ports and primary Anastomosis was carried out uneventfully. Postoperative period was uneventful and is doing well at 8 years with micronutrient therapy and laxatives intermittently for associated hypo-ganglionosis.

Case 3

A 36 hours old male term infant weighing 3200 mg. At the 2nd day of life was admitted at local community hospital for failure to pass meconium, no bowel movements despite feeding and no response to glycerin suppository and persistent non-bilious vomiting. Physical examination revealed a distended abdomen, no associated anomalies were found.

Abdominal ultrasound showed normal pyloric canal length of 12 mm and thickness of 2mm and rest of the ultrasound was normal. Chest radiograph showed the tip of the feeding tube was in the stomach. Abdominal radiograph revealed a large dilated viscus lying the right of midline with some dilated loops of further bowel, probably representing an obstructed small bowel and no distal or rectal gas suggesting low bowel obstruction such as bowel atresia, meconium ileus or long segment Hirschsprung's disease (Fig 3 A). He was resuscitated and transferred to our regional Neonatal Care Unit.

A water-soluble enema was performed. The colon was displaced to the left side of the abdomen by a large air-filled loop of bowel seen within the right iliac fossa. No meconium, air passed or anorectum gripping the catheter during the rectal examination. Contrast then passed through rectum, sigmoid colon, the transverse and hepatic flexure without any transition zone. No contrast could be passed further despite repeated attempts even using hand pressure. No meconium was identified during the examination. Ascending colonic atresia with dilatation of the proximal caecum and small bowel was very likely (Fig 3B).

Minimal Invasive Surgery via periumbilical incision revealed upper ascending Colon Atresia type 3 with close loop dilatation of proximal ascending colon and cecum

with competent ileocecal valve and diffuse secondary small bowel dilatation. Resection of both dilated end and atretic ends with ceco-colic anti-mesenteric tapering ceco-coloplasty and end to end Anastomosis was carried out uneventfully. Rectal suction biopsy and excised specimen at surgery showed normal ganglion cells with no evidence of Hirschsprung's disease. Immediate postoperative period was uneventful and discharged home after 6 days.

Postoperatively, a water-soluble contrast was injected through a rectal tube. There is free flow of contrast through the distal colon, which has small caliber into the proximal part of the transverse colon, at the site of the surgical anastomosis of corrected Colonic Atresia. The contrast accumulated gradually in a markedly distended ascending colon and cecum and in turn, refluxed into the terminal ileum through incompetent ileocecal valve. The progressively distended ascending colon and cecum could be related to partial obstruction at the site of surgical Anastomosis or Dysmotility.

However, in few months' time, represented with gradually increasing abdominal distention, anemia and failure to thrive and nasogastric tube culture grew candida. Abdominal ultrasound suggested multiple dilated loops of bowel seen throughout the abdomen. Abdominal radiograph showed small and right sided large bowel with gas in the distal colon suggestive of partial obstruction (Fig 3C). Contrast studies showed partial functional obstruction with hugely dilated previously tapered ceco-coloplasty segment, patent anastomosis and no distal obstruction (Fig 3D).

He underwent re-explanation through the same periumbilical scar cutting incision and the hugely dilated right colon was resected with partial subtotal excision of the cecum, preservation of the appendix and ileocecal valve, appendicostomy with a silastic tube with multiple holes was passed into terminal ileum to decompress it and appendix tip was stitched with lateral abdominal peritoneum to exteriorize and the silastic tube appendico-ceco-ileostomy temporary proximal decompression

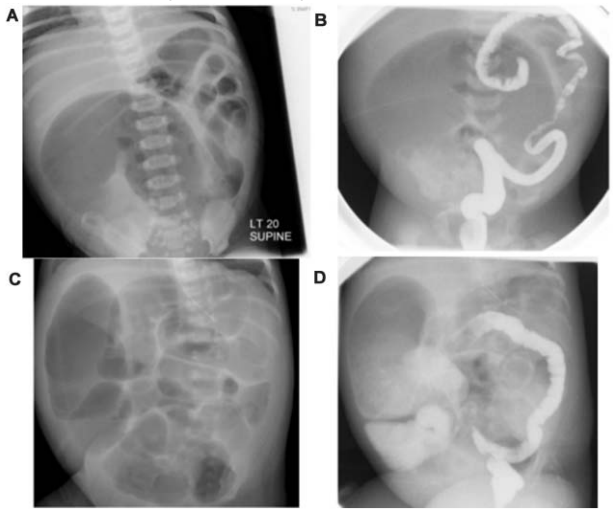


Fig 3 — Pre and post operative Plain abdominal radiograph and contrast enema of case 3

followed by ceco-colic end to end anastomosis was carried out (Fig.4A). The postoperative period was uneventful and the silastic tube was removed after 7 days which closed itself (Fig 4B). He is doing well at 6 years follow up.

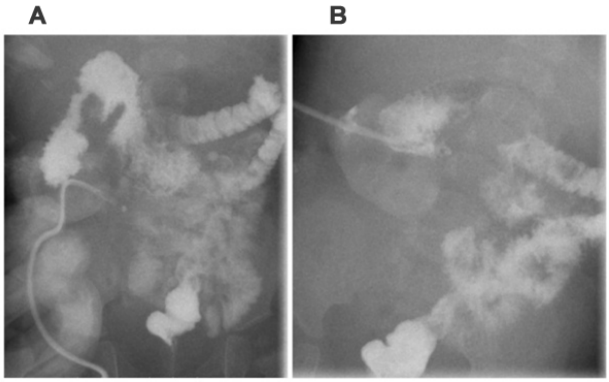


Fig 4 — Follow up postoperative prograde contrast via appendicostomy tube in Case 3

Case 4

Term baby, 38 weeks of gestational age was born with 2578-gram weight to a diabetic mother, who was positive for HIV for 18 years of age, currently with plasma viral levels undetectable.

On the 1st day of life, the baby presented with milky vomiting, has not passed meconium despite glycerin suppository with a normal size and site of the anus and per rectal examination did not reveal any gripping finger or gush of gas or fecal matter suggestive of Hirschsprung's disease. In a couple of hours, clinical deterioration and abdominal distension was noticed and the abdominal radiograph and a decubitus view suggested multiple air fluid levels and step ladder pattern suggestive of low intestinal obstruction (Fig 5 A and B). Repeat radiographs at 6 hours suggested increased distention and no gas in the distal colon or rectum with dilated proximal bowel loops and air fluid levels (Figs 5 C & D).

The baby was quite ill and resuscitated. Once stabilized, the baby was taken to operation theatre and an exploratory laparotomy with the diagnosis of low intestinal obstruction.

At the exploration, stomach, duodenum and jejunum were normal. A segment of terminal ileum starting at about 10 cm of the ileo-cecal valve, was hugely dilated, extending on to the caecum that also was very large and ending in a atretic segment as a blind pouch in the region of transition between caecum and right colon with normal appendix. The rest of the colon was not present except for the rectosigmoid stump in the pelvis supplied by superior rectal vessels (Fig 6, A-D). The middle colic vessels and the inferior mesenteric vessels were missing.

An Ileostomy was performed. The patient started to receive feeds on the 3rd day postoperative, increasing slightly which were all tolerated very nicely while stoma was functioning well. Baby was finally discharged home

after 14 days. The plan is to slow down the motility of proximal functioning gut using loperamide and increase micronutrient absorption and lengthen the bowel, rectal suction biopsy to exclude Hirschsprung's disease or other congenital motility disorders and volume expansion of the distal rectosigmoid stump to lengthen and widen and allow Anastomosis and gain extra colon surface for absorption of water and electrolytes thus reducing diarrhea after restoring the continuity with ileo-colic anastomosis at a later date.

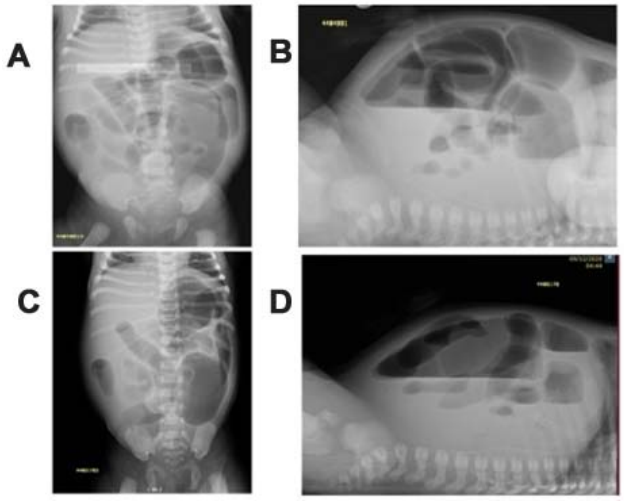


Fig 5 — Plain and decubitus view of abdominal radiographs after birth-Case 4

Case 5

Male term baby was born with 38 weeks of gestational age and had no pre-natal abnormalities on scans and no perinatal problems. Breast feeding was started but on the second day of life was brought to intensive care due to abdominal distension and bilious vomiting, The anus was at normal site and of normal size and no meconium found on the rectal examination. Babygram showed dilated stomach and bowel gas in the abdomen and no gas in the rectum or pelvis (Fig 7A).

The abdominal circumference increased rapidly over the next hours and a contrast enema showed microcolon and cut off sign at rectosigmoid junction with no meconium but small mucoid white plug came out suggestive of Colonic Atresia (Fig 7 B).

At laparotomy Global dilatation of the small intestines as well as the entire colon until the transition between the left colon and the sigmoid, where a type I atresia was present (Fig 7 C-F). A diverting loop stoma was performed proximally to the site of the atresia. Postoperative period was uneventful and a suction rectal biopsy to exclude associated Hirschsprung's disease followed by volume tissue expansion of the distal stump using saline is planned before closure of the stoma.

Discussion

We strongly believe in ileo-cecal valve, appendix and colonic preservation in atresia where associated short

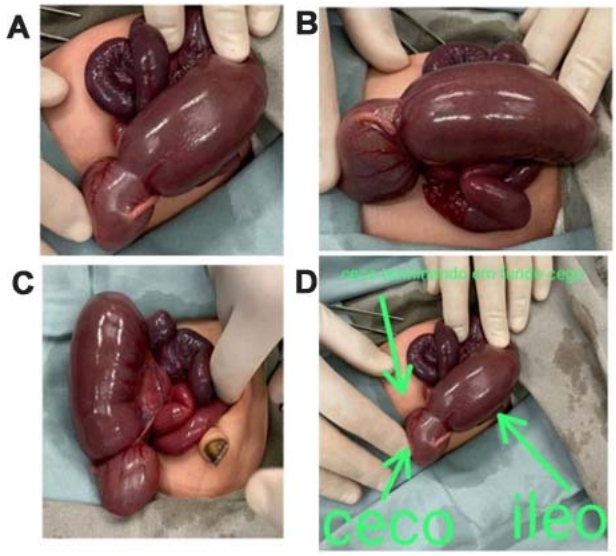


Fig 6 — Operative photographs, note blind ending cecum and dilated terminal ileum with absent colon and colorectal stump-Case 4

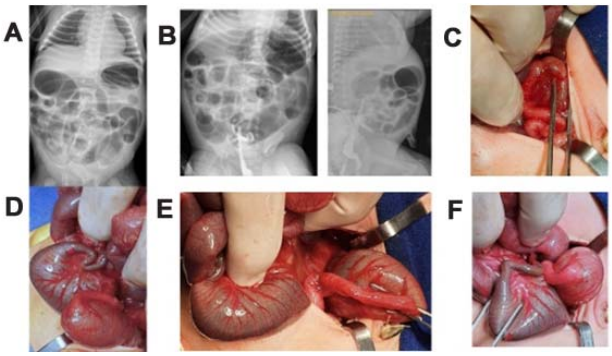


Fig 7 — radiological and operative findings in Case 5

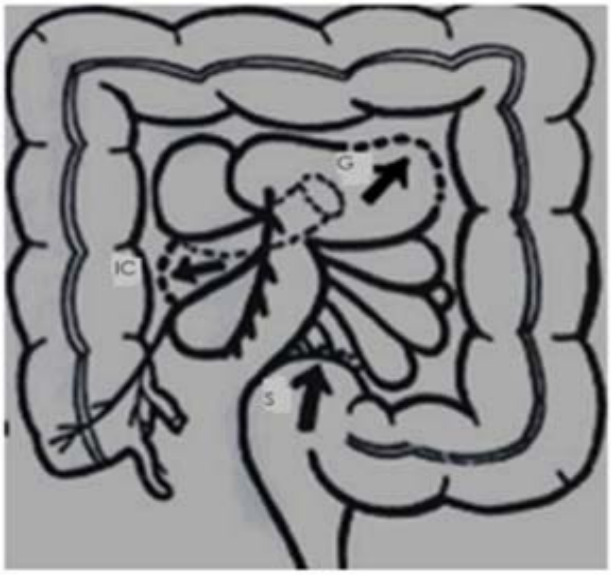


Fig 8 — Colonic blood supply with three weak points in the marginal arterial arcade

bowel and variable length of bowel may have been absorbed and missing, in advanced pediatric intussusceptions and adult population whenever feasible[12-15]. Rupture of intussuscepiens and dual transanal and transperitoneal prolapse of intussusception was first reported in 1982 as an unusual and rare complication[16]. These cases came mainly from north and west of India and an anecdotal case was reported from Europe and a plausible explanation that it is due to the prolonged and constant pressure of the intussusception in the antimesenteric part of the intussuscepiens, which is already stretched in a convex arch did not appear altogether satisfactory[16-19]. However, it was only later when we observed venous thrombosis of the intussuscepiens and marginal arterial arcade defects in a similar case in a West African infant presenting late, the vascular etiology of this phenomenon was very clear[19].

The most likely sites of Colonic Atresia would be where the Anastomosis of the marginal artery is least effective. The marginal artery does not link up certain vessels in a rare congenital anomaly of colonic blood supply as shown in figure 8, thus forming three weak points in the arcade[20]. If the artery supplying the area of the atresia has been affected beyond the neighboring branch at these sites, there is little chance of an effective collateral circulation being established[21-22]. It is very rare to involve multiple points in the marginal arterial arcade which happened in our fourth case as an extreme rarity. Colonic Atresia cannot be diagnosed prenatally, usually not suspected as very rare and pre-operative diagnosis requires contrast enema.

There is an association of Hirschsprung's disease or its variants in the colon distal to the site of atresia due to arrest of migration of craniocaudal migration of ganglion cells[23]. It is therefore, suggested that all cases of Colonic Atresia must be biopsied from distal part of the colon as well as rectum to rule out Hirschsprung's disease or its variants as an associated anomaly. Further detailed studies are required based on histology and immunohistochemistry of intestine to decide the level of resection of the proximal dilated colon and distal colon to achieve early bowel activity and reduce the morbidity[24].

We have previously reported modified classification of intestinal and biliary atresia-cystic malformations[25-27] and now we suggest a modified spectrum of atresia-stenosis-cystic malformation classification of these lesions and we believe that it is a continuation of the spectrum depending on the altered vascular supply of the segment (Table 1). We have made

Table 1 — Atresia-segmental dilatation-cystic malformation spectrum classification

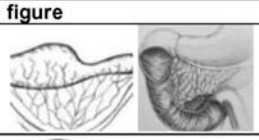
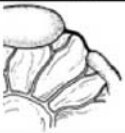
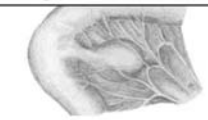
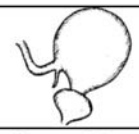
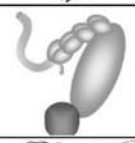
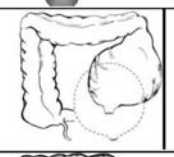
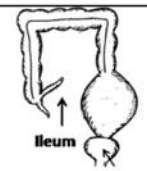
Type	Description	figure
Type 0	Stenosis/Web	
Type I	Atresia with a mucosal defect	
Type II	Fibrous cord connecting the atretic end	
Type IIIa	Atresia with a V-shaped mesenteric gap defect	
Type IIIb	Apple-peel deformity	
Type IV	Multiple atresia	
Diverticulum	Part of the wall protrusion with preservation of embryonic blood supply	
Fusiform dilatation /Segmental dilatation/stenosis	A segment of bowel having stenosis or dilatation as a result of altered blood supply	
Cystic malformation Type!	Normal colon absent, ileum enters the pouch	
Type II	Subtotal (except Caecum and part of ascending colon) colonic cystic dilatation	
Type III	Left colonic cystic dilatation	
Type IV	Localised Rectosigmoid colonic cyst	
Type V	Multiple Cystic dilatations with normal segment of colon interposed	

Table 2 — *Prognostic subclassification of atresia-segmental dilatation-cystic malformation spectrum*

Type	Lesion	Prognosis
A	Isolated	excellent
B	Multiple single system	good
C	Multisystem anomalies	poor
D	Syndromic/Genetic	worst

easy subclassification of these lesions based on prognosis of these lesions into subdivisions ranging from A to D (Table 2).

Conclusion

Left Transverse Colostomy in rectosigmoid atresia at Sudeck point is more convenient than the Right Transverse Colostomy as it reduces colostomy associated diarrhea and morbidity and mortality as compared to right sided Transverse Colostomy. Hydrodistension of the distal rectosigmoid stump is a very good tissue expansion technique which allows the distal colon to be of good caliber reducing disparity between proximal and distal segments and coco-colostomy at a later date will be easier[28]. Although our experience with coloplasty from dilated colon in pouch colon syndrome suggest that the tube coloplasty may not be long lasting but certainly it provides a temporary conduit and allows initial growth and development and it allowed preservation of appendix and ileocecal valve in our third case. In conclusion, we believe that even if prenatal diagnosis is not possible in Colonic Atresia, high index of suspicion and immediate plain radiograph and contrast enema allows prompt diagnosis by differentiating with other distal intestinal obstruction such as Hirschsprung's disease, meconium ileus, left colon syndrome, etc and appropriate management provides excellent prognosis.

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