



Autopsy and Case Reports

ISSN: 2236-1960

Hospital Universitário da Universidade de São Paulo

Krishnamurthy, Kritika; El Hussein, Siba; Omarzai, Yumna  
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Autopsy and Case Reports, vol. 8, no. 3, e2018037, 2018, July-September  
Hospital Universitário da Universidade de São Paulo

DOI: <https://doi.org/10.4322/acr.2018.037>

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## The lethal twist – a story of unspoken pain: small intestinal volvulus in cerebral palsy

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**How to cite:** Krishnamurthy K., El Hussein S., Omarzai Y. The lethal twist – a story of unspoken pain: small intestinal volvulus in cerebral palsy. Autops Case Report [Internet]. 2018;08(3):e2018037. <https://doi.org/10.4322/acr.2018.037>

### ABSTRACT

Small intestinal volvulus (SBV) is the abnormal twisting of bowel around the axis of its mesentery, leading to obstruction and vascular compromise, resulting in bowel ischemia and necrosis which are life-threatening. Risk factors include malformation, malrotation, and adhesions. Its rare incidence and vague clinical presentation make it a difficult diagnosis, more so in a nonverbal patient who cannot express his pain, which is the first and most prominent symptom. Studies suggest an increased frequency of intestinal obstruction in cerebral palsy patients. There are no reported cases of small intestinal volvulus in association with cerebral palsy. We present a case of a 21-year-old man with severe cerebral palsy and kyphoscoliosis. The patient presented to the emergency room with respiratory distress and abdominal distension. An acute abdomen was noted. Abdominal X-rays revealed gas patterns suggestive of small intestinal obstruction. The patient rapidly deteriorated, and resuscitation attempts were unsuccessful. Autopsy revealed peritoneal cavity filled with extensively dilated and thin-walled loops of small intestine. Twisting of the small intestine, showing 360° rotation around the mesenteric root in a clockwise manner at two separate sites, was noted. On bowel dissection, mucosal folds were absent, and mucosa was green with patchy areas of hemorrhage consistent with ischemic necrosis. There was no evidence of any malformation, malrotation or adhesions. Small intestinal volvulus is a rare entity with a nonspecific clinical presentation that poses a diagnostic challenge. This autopsy highlights the need to maintain a high index of suspicion for small intestinal volvulus in cases of bowel obstruction in cerebral palsy patients to expedite surgery and prevent mortality. The primary caregivers of non-verbal cerebral palsy patients living outside of healthcare facilities need to be trained in recognition of life-threatening medical emergencies such as gastrointestinal obstruction and seek emergent attention at the earliest to prevent treatment delays.

### Keywords

Intestine, Small; Intestinal Volvulus; Cerebral Palsy; Kyphoscoliosis.

### CASE REPORT

We present a case of a 21-year-old man with severe cerebral palsy, kyphoscoliosis, asthma and epilepsy, who was being fed through a percutaneous endoscopic gastrostomy (PEG) tube for the past several years. The patient had severe cognitive impairment

and was nonverbal, and was brought in with severe respiratory distress and abdominal distension. The primary caregiver, his mother, complained of being unable to push food through the PEG tube for the previous two days. The suction of the PEG tube

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was performed, with no improvement in symptoms. Abdominal X-rays revealed abnormal gas patterns and air-fluid levels suggestive of small intestinal obstruction. Intestinal obstruction was suspected; however, surgical intervention could not be initiated because the patient's condition deteriorated rapidly. Resuscitation attempts were unsuccessful, and the patient expired within 3 hours of initial presentation.

## AUTOPSY FINDINGS

On external examination, the corpse was that of a poorly developed and poorly nourished man with severe scoliosis and musculoskeletal sequelae consistent with cerebral palsy. The abdomen was markedly distended with green discoloration of the overlying skin (Figure 1A). The PEG tube was located just above the umbilicus and was full of straw-colored fluid. The peritoneal cavity contained 250 ml of serosanguinous fluid and was filled with extensively dilated loops of small intestine with green discoloration (Figure 1B).

Twisting of the small intestine, showing 360° rotation around the mesenteric root in a clockwise manner at two separate sites, was noted (Figure 2A). The mesenteric veins were engorged (Figure 2B).

Dissection of the distended and thin-walled small bowel revealed changes consistent with ischemic necrosis including green mucosal surfaces, absence of mucosal folds and patchy areas of hemorrhage. There was no other evidence of any developmental malformation, malrotation or adhesions. Histopathological examination of the dilated segments revealed focal transmural hemorrhage (Figure 3A) and necrosis along with diffuse thinning of epithelium and muscularis propria (Figure 3B).

## DISCUSSION

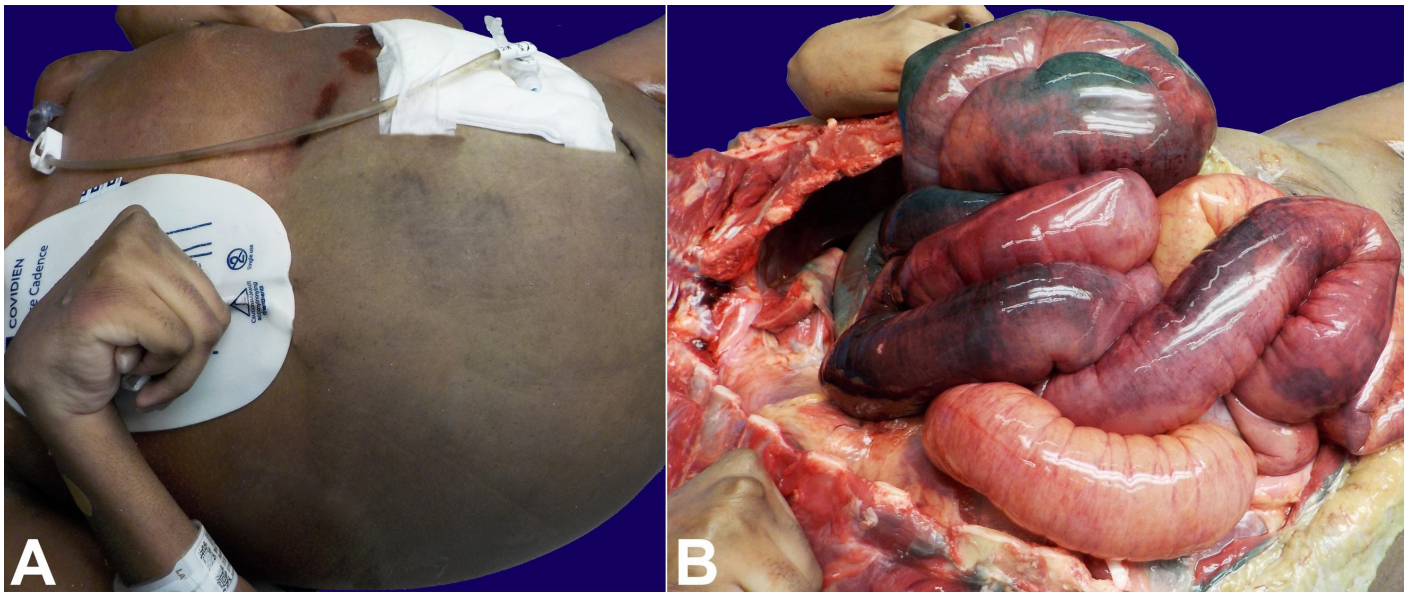
Cerebral palsy is associated with an increased risk for mortality at an earlier mean age than the general population.<sup>1</sup> The most frequently cited causes of death are related to the circulatory and respiratory systems.<sup>2</sup> However, clinically significant gastrointestinal abnormalities such as defective swallowing reflex, delayed gastric emptying, chronic constipation, and intestinal obstruction especially small intestinal

obstruction are known complications of cerebral palsy that result in increased morbidity and can contribute to an increased risk for mortality.<sup>3,4</sup> While gastric and colonic volvulus have been more frequently reported in cerebral palsy patients, small intestinal volvulus is a life-threatening, albeit rare condition that needs to be recognized quickly, and treated urgently in this patient population.<sup>5,6</sup>

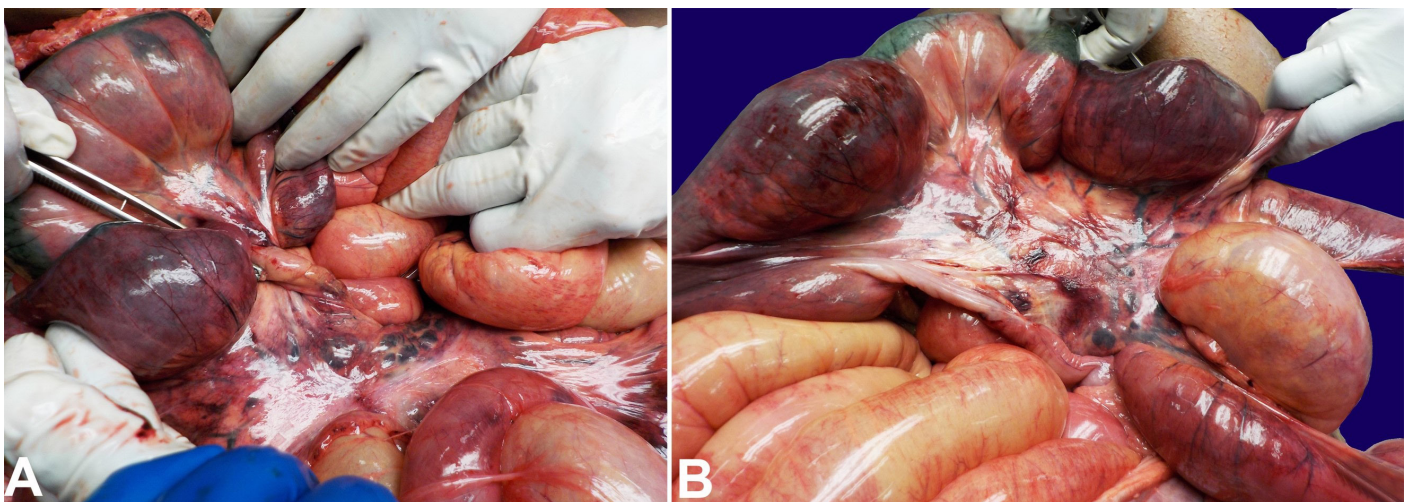
SBV, one of the etiologies of small intestinal obstruction, is the abnormal twisting of a loop of small intestine around the axis of its mesentery, leading to mechanical obstruction and vascular compromise. The torsional strain around the mesenteric vasculature results in the strangulation of the blood supply leading to bowel ischemia and bowel necrosis which are life-threatening. The clinical presentation is that of acute abdomen with nonspecific symptoms of nausea, vomiting, and abdominal pain. Thus, this entity is hard to recognize especially in a nonverbal patient who cannot express his pain, which is the first and most prominent symptom. Cerebral palsy can lead to severe cognitive impairment and lack of effective communication. The primary caregiver for such patients should be counseled about the increased incidence of gastrointestinal obstruction in their patient and trained to recognize and seek emergent attention for the same. The importance of this is exemplified in our case, where the caregiver failed to suspect obstruction despite the inability to push feeds through the gastrostomy tube resulting in treatment delay and, subsequently death.

Small bowel volvulus is infrequently encountered and comprises 1-6% of cases of small bowel obstruction cases in the adult population.<sup>7</sup> Small bowel volvulus is classified as primary and secondary types.<sup>8</sup> Primary SBV occurs in the absence of any anatomical abnormality of the bowel and its etiology is poorly understood. Multiple studies investigating primary small bowel volvulus suggest that this entity is caused by a combination of factors.<sup>9,10</sup> A long mesenteric length associated with a short mesenteric root would permit abnormal mobility of the small bowel or a segment of it. An important precipitant of twisting might include abrupt changes in dietary habits such as ingestion of bulky food in large amounts after long intervals of fasting. It is thought that the sudden filling and overloading of an empty bowel by ingestion of a poorly digestible and voluminous meal may induce

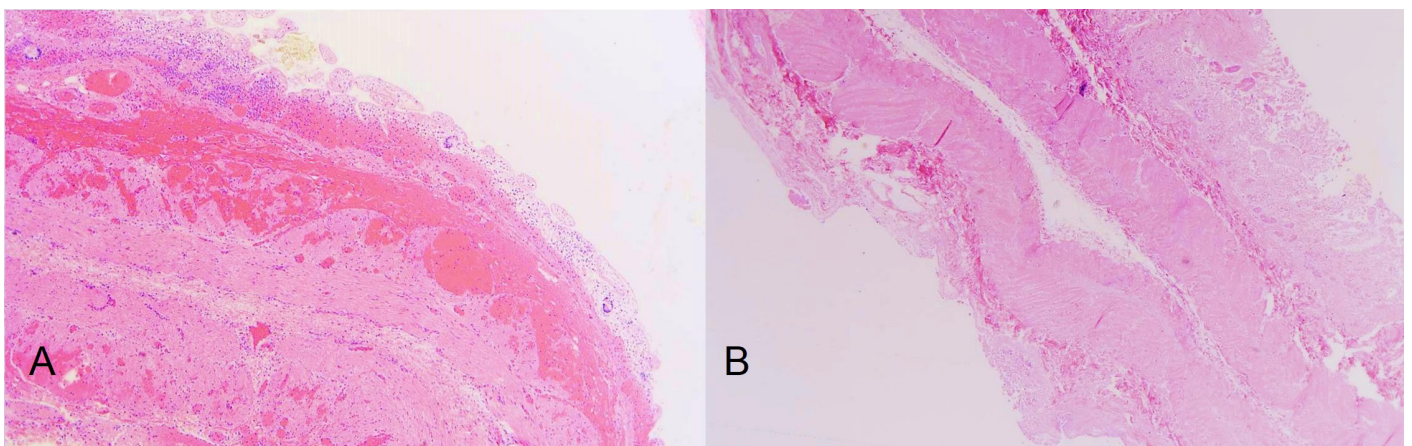




**Figure 1. A** – Gross examination of the corpse showing markedly distended abdomen with green discoloration of overlying the skin and PEG tube in situ; **B** – Macroscopic examination of the abdominal viscera with extensively dilated thin-walled loops of small intestine with green discoloration.



**Figure 2. A** – Macroscopic view of the small bowels with a clockwise twisting of the small intestine around the mesenteric root; **B** – Note the engorged mesenteric veins.



**Figure 3. A** – Focal transmural hemorrhage; **B** – Diffuse thinning and necrosis of the intestinal wall.



forceful bowel peristalsis, resulting in the initiation of a volvulus. There are no studies examining the role of specific dietary habits or requirements, such as tube feeding in our case, in the causation or progression of small bowel volvulus.

Secondary SBV occurs in the presence of an acquired condition, such as congenital malrotation, anatomic abnormalities such as atresia or mesenteric defect, congenital fibrous bands, postoperative adhesions, tumors, pregnancy, and diverticular disease.<sup>11</sup> The mechanism of torsion is related to abnormal fixation points in these cases.

Small bowel obstruction generally presents with severe diffuse or vague abdominal pain, nausea, vomiting accompanied by constipation and a lack of normal flatulence. The diagnosis of small bowel volvulus is very difficult to make because there are no distinct pathognomonic clinical signs or symptoms that distinguish between simple and strangulating obstruction. Conventional radiographs show abnormal intraluminal gas patterns; however, these findings are non-specific and fail to establish small intestinal volvulus as the cause of the small intestinal obstruction.<sup>12</sup> Even a normal appearing abdominal film does not eliminate the possibility of a small bowel volvulus. CT scans may reveal torsion of the loops of small bowel around the mesenteric vessels and mesentery known as the “whirl sign”.<sup>13</sup> However, the sensitivity and specificity of the whirl sign for a diagnosis of SBV is variable due to operator subjectivity and other confounding factors.<sup>12</sup>

## CONCLUSION

Although small bowel volvulus is a rare entity, it should be included in the differential diagnosis of bowel obstruction in patients with cerebral palsy. It is an easily unrecognized occurrence in a non-verbal patient. Its infrequent incidence, along with the vague clinical presentation and subtle radiologic findings, make it a difficult diagnosis. The subsequent delay in surgical intervention results in bowel necrosis that can be rapidly fatal. This autopsy highlights the need to maintain a high index of suspicion for small intestinal volvulus in cases of bowel obstruction in cerebral palsy patients to expedite surgical intervention and prevent mortality. The primary caregiver in such cases should be well trained to recognize the earliest signs

of life-threatening emergencies such as gastrointestinal obstruction and seek emergent attention preventing treatment delays and death.

The authors retain an informed consent signed by the relatives for the autopsy performance and the manuscript was approved by the Institutional Ethics Committee.

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**Authors contributions:** Krishnamurthy K. and El Hussein S. performed the autopsy and the photography. Krishnamurthy K. and Omarzai Y. did the microscopic examination, finalized the autopsy and worked on the manuscript. All the authors read and approved the manuscript.

**Conflict of interest:** None

**Financial support:** None

**Submitted on:** June 02<sup>nd</sup>, 2018

**Accepted on:** June 27<sup>th</sup>, 2018

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