# Idiopathic Megacolon in an Adolescent Girl: a Case Report on a Rare Disease

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**Research Article**

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# Abstract

**Introduction:** Idiopathic Megacolon is a rare condition where there is persistent dilatation of colon in the absence of identifiable cause. Symptoms start as early as in childhood or in adulthood. Colectomy have shown to have high success rate in patient with idiopathic megacolon. We reported a case of an adolescent girl with idiopathic megacolon that was successfully treated with colectomy.

**Presentation of case:** A 15 years old girl presented with a complaint of abdominal pain associated with gradual abdominal distension for 1 week duration, not passing flatus and had multiple episodes of vomiting. She was diagnosed to have sigmoid volvulus and underwent endoscopy decompression, however symptoms recurred. Segmental colectomy was performed and she had an uneventful recovery.

**Discussion:** Patient presented with acute intestinal obstruction attributed to sigmoid volvulus with a history of constipation. A diagnostic dilemma between Hirschsprung's disease with other causes of megacolon occurred as these diseases have similar presenting symptoms. A confirm histopathology of presence of ganglionic cells within the plexus exclude the diagnosis of Hirschsprung's disease hence the diagnosis of Idiopathic Megacolon was made. Segmental colectomy give good result in relieving patient symptoms of abdominal pain and constipation and one of the recommended surgical options in treating Idiopathic Megacolon.

**Conclusion:** Idiopathic megacolon is a rare disease and for a long time has been a disease of exclusion. Rectal biopsy is important to exclude the diagnosis of Hirschsprung's Disease. There are wide variety of surgical treatment available in treating Idiopathic megacolon. Segmental Colectomy have shown good success rate.


# 1. Introduction

Idiopathic Megacolon is a rare condition, describe as a condition where there is persistent dilatation of colon in the absence of identifiable causes. It can affect both sexes equally [1] and symptoms may start as early as in childhood or in adulthood. The exact mechanism is unknown however it is postulated that there are abnormalities in the extrinsic and enteric nervous system, intestinal smooth muscle or neurotransmitter which cause colon dilatation and impaired colonic motility [2]. Atrophy of the collagenous tendinous connective tissue membrane of the myenteric plexus and tendinous collagen fiber net of the muscularis propria allow for unlimited distension of the colon [2]. Constipation, abdominal distension and pain are the common presentations in adults meanwhile fecal impaction and soiling are more common in children [3]. Colectomy have shown to have high success rate in patient with idiopathic megacolon where there are relief of symptoms of abdominal pain and constipation and devoid requirement on laxatives [4]. Here we reported a case of an adolescent girl with idiopathic megacolon that was successfully treated with colectomy.

# 2. Case Presentation

A 15 years old girl with no known underlying co-morbidity, presented with a complaint of abdominal pain associated with gradual abdominal distension for 1 week duration. She was not passing flatus and had multiple episodes of vomiting at home. She gave a history of multiple similar complaints before however symptoms was self-limiting after a few days without any treatment. Apart from that, she also had a history of constipation since early childhood but did not seek treatment. Her usual bowel habit was 3x per week. She had difficulty to empty the bowel and also had the need to strain to pass motion. Two weeks prior to her presentation to us, she was admitted to a different hospital with similar complaints. Abdominal x-ray was performed which showed dilated large bowel with coffee bean appearance over the sigmoid region [Figure 1]. A diagnosis of sigmoid volvulus was made. She underwent endoscopy decompression, however the symptoms recurred after three days at home. At our center, clinical examination revealed that she was dehydrated, tachycardia however normotensive and afebrile. Abdomen was grossly distended and tender with hyperactive bowel sound. Rectal examination showed empty rectum. She had normal hemoglobin level and leucocytes count. Renal functions were normal. Abdominal x-ray revealed dilated large bowel and absent of rectal gas [Figure 2]. She was treated conservatively as the abdominal x-ray did not show a diagnosis of sigmoid volvulus. Her condition was not improving despite conservative treatment and there was increasing of abdominal distension. She started to feel more discomfort due to an increasing abdominal distension. She underwent an emergency laparotomy and it revealed that the sigmoid colon till the descending colon was grossly dilated. The transverse colon was normal. Segmental colectomy was performed involving the resection of the grossly dilated sigmoid and descending colon. The distal end was closed at the upper rectum and the proximal end was brought out as end transverse colostomy. Rectal biopsy was also performed to exclude Hirschprung's disease on her. She had uneventful recovery and was discharged after one week. Histopathology revealed hypertrophy of the muscularis mucosa, otherwise there were normal number of ganglia in the myenteric plexus and no hypertrophic nerve bundle. Rectal biopsy revealed there were sparse, small myenteric ganglia with hypertrophy of muscularis mucosae. There were no hypertrophic nerve bundles seen. A diagnosis of Idiopathic Megacolon was made on her. She had recovered well with the abdominal symptoms resolved completely. She underwent re-laparotomy, re-anastomosis of the transverse colon to the upper rectum after eight weeks from the first surgery. She again had an uneventful recovery and discharged well. A few follow up visits subsequently were unremarkable.

# 3. Discussion

Megacolon can be defined as increased in bowel length and diameter without involvement or insignificant involvement of rectum [5]. The pathology can be further subdivided according to its etiology into congenital (Hirschsprung's disease), acquired and idiopathic megacolon thus idiopathic megacolon is described as megacolon without obvious etiology [6].

The incidence of idiopathic megacolon is unknown and it affects both gender equally [4]. As a result of its rarity, the diagnosis of idiopathic megacolon is not uniform and its greatly depending on the individual attending surgeon. A comprehensive diagnostic criteria based on a systematic review was proposed by Cuda et al. which consist of 3 criteria [5]:

1. Intact anorectal inhibitory reflex or exclusion of organic pathology by rectal biopsy.
2. Radiological imaging (Abdominal X-ray or Barium enema) that showed 10cm diameter dilatation of the sigmoid colon.
3. Presenting symptoms of gas distress, abdominal pain, distension and constipation.

The patient presented with acute intestinal obstruction attributed to sigmoid volvulus with a history of constipation. Her abdominal x-ray showed megacolon with absent of rectal gas. A diagnostic dilemma between Hirschsprung's disease with other causes of megacolon occurred as these diseases have almost similar presenting symptoms. Symptoms of constipation, abdominal pain with distension and even intestinal obstruction with sigmoid volvulus [8] may occurred in any type of megacolon. Rectal biopsy has been recognized as the important diagnostic tool with 95% accuracy in Hirschsprung's disease diagnosis [7]. Hence in this patient, rectal biopsy was performed and the diagnosis of Hirschsprung's disease was excluded.

To date, there is no published guidelines on management of Idiopathic Megacolon. This result in wide variety of surgical treatment adopted by the attending surgeon. This patient underwent segmental resection of the dilated colon with the proximal end was brought out as end stoma. In one systematic review, it recommended the procedure of choice for idiopathic megacolon were subtotal colectomy, segmental bowel resection and fecal diversion by means of creating diversion stoma [4]. Of these surgical intervention, subtotal colectomy was the treatment that imposed the highest positive outcomes however still having significant morbidity involving intestinal obstruction [4]. Segmental resection follows with successful rate of 48.4% but came with risk of recurrent symptoms [4].

# 4. Conclusion

Idiopathic megacolon is a rare disease and for a long time has been a disease of exclusion. A confirm histopathology of presence of ganglionic cells within the plexus exclude the diagnosis of Hirschsprung's disease hence the diagnosis of Idiopathic Megacolon was made. This is crucial to achieve a definite diagnosis in order to offer a proper treatment for the patient. Many studies have shown colectomy offers a good result with minimal complications in the treatment of Idiopathic Megacolon. Segmental colectomy give good result in relieving patients symptoms of abdominal pain and constipation and one of the recommended surgical options in treating Idiopathic Megacolon.

# Declarations

### Competing interests

Authors have declared that no competing interests exist.

### Authors' Contributions

All authors contributed equally in the manuscript.

### ETHICS APPROVAL

Not Applicable

### Consent

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

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## Figures

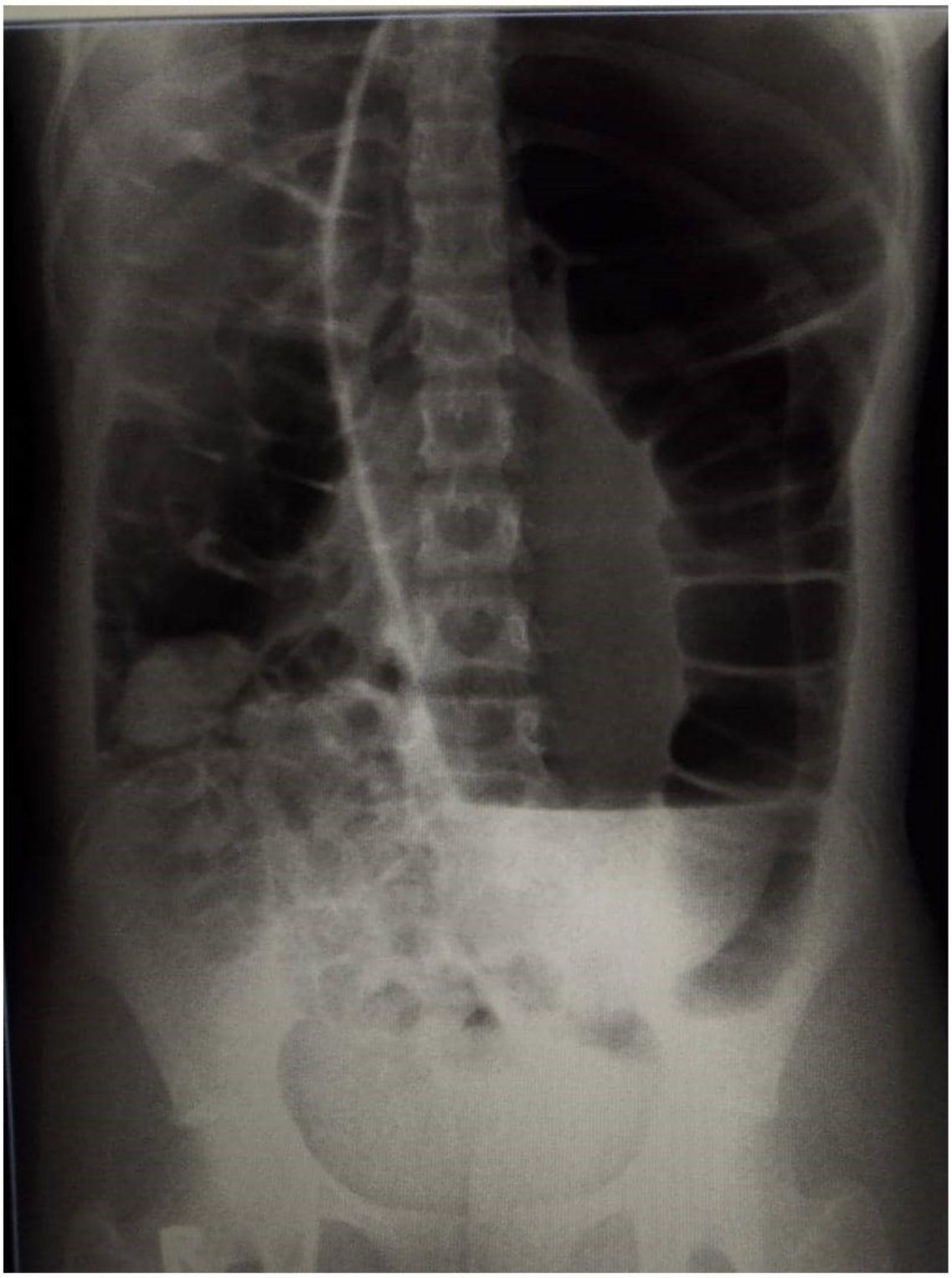

**Figure 1**

Abdominal x-ray showed dilated large bowel with coffee bean appearance over sigmoid region to suggest sigmoid volvulus.

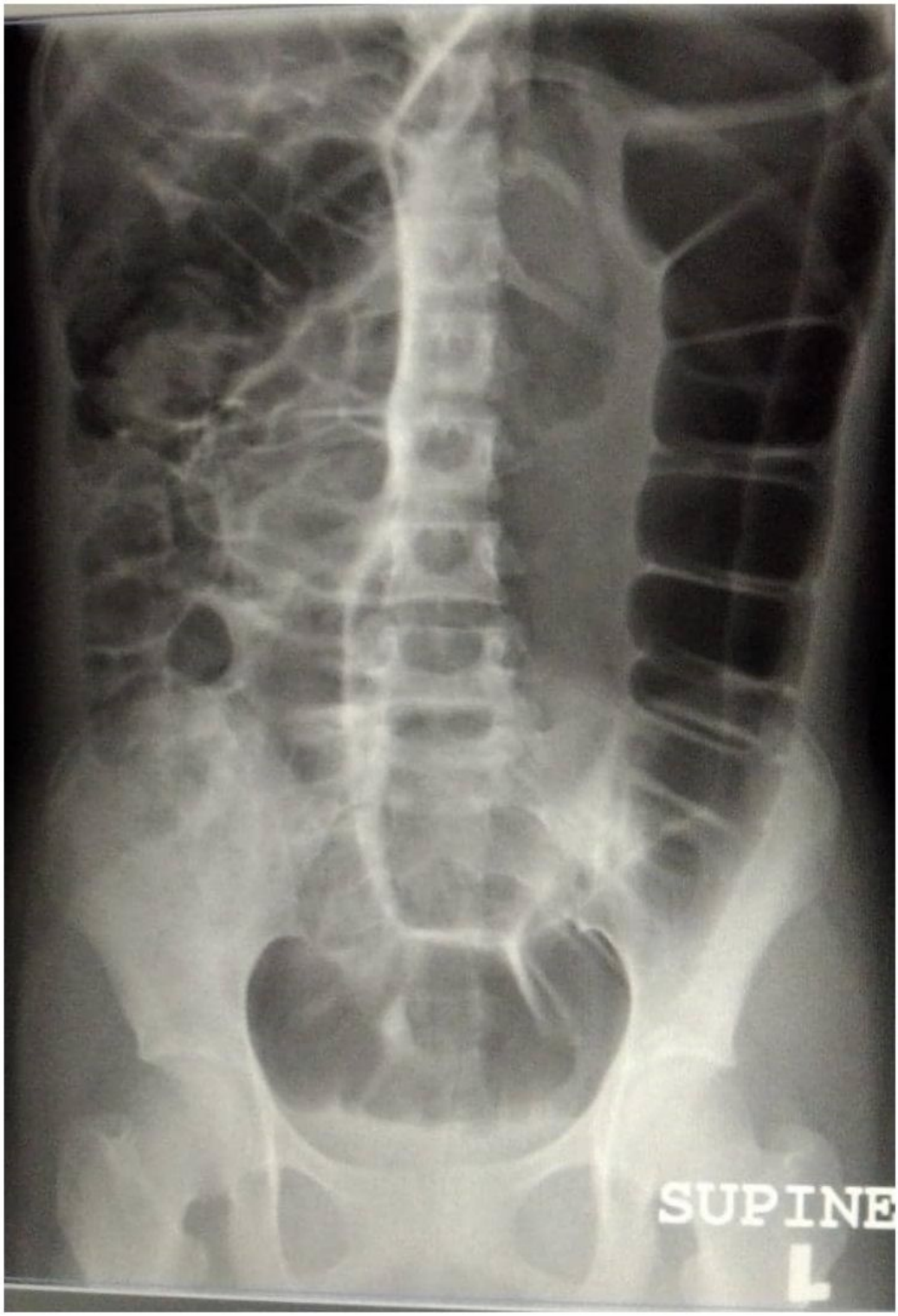


**Figure 2**

Abdominal X-ray showed grossly dilated large bowel with no rectal gas seen.

## Supplementary Files

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