Adult Hirschsprung's Disease in 20 Years Old Female: Report of Case
Ibrahim Salih Elkheir1, Hozifa Mohammed Ali Abd Elmaged2, Sara Gamareldein Abdalla3, Tagwa Faisal Mohamed Alsadig4

1Pediatric Surgeon, Khartoum Bahri Teaching Hospital; Associate professor at Alzaeim Alzahari University Sudan
2Orthopedic Resident, Khartoum Bahri Teaching Hospital; teaching assistant at Alzaeim Alazhari University Sudan
3Pediatric Resident SMSB
4MBBS Alzaeim Alazhari University-Sudan

*Corresponding author: Ibrahim Salih Elkheir
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Abstract
Hirschsprung’s Disease is a congenital disease of the colon that rare seen in adult and usually misdiagnosed or undiagnosed. And characterized by the lack of ganglion cells of the sub mucosal (Meissner) and myentric (Auerbach) neural plexuses in the affected segment of the bowel. We present a 20 years female who had absolute constipation with abdominal distention and vomiting and had history of delayed of passage of meconium with recurrent episode of constipation since childhood that required recurrent hospital admission. The diagnosis was made upon the history and rectal biopsy that showed there's absence of ganglion cells in myentric (Auerbach) and sum-mucosal (Meissner) plexus in the distal colon which is typical to the classical form of hirschsprung’s disease. She treated firstly with sigmoid colostomy followed by pull through operation after she was returned back to hospital at 25 years old with no complication postoperatively. So Hirschsprung’s disease should be considered in the causes of recurrent constipation in any adult case.

Keywords: Hirschsprung’s Disease, Adult, constipation, good outcome.

INTRODUCTION
Is a congenital disorders characterized by lack of ganglion cells in the sub mucosal and myentric neural plexus mainly in the rectosigmoid or the rectum. Is commonly seen in infant and newborn but is rare seen in adult.

Common symptoms include delayed passage of meconium, recurrent constipation and failure to thrive in early childhood [1, 2].

However, some patients with mild symptoms may go undiagnosed into adulthood, likely because the colonic region proximal to the distally obstructed segment assumes a compensatory role [3,4]. But at end the dilated colon is no longer able to propel the faces distally.

Adult commonly present with recurrent constipation or fecal retention and treated by enema to relieve the symptoms.

Diagnosis is confirmed when there is aganglionosis in the rectal biopsy. Here we report a 20 years female presented with absolute constipation for 4 days with abdominal distention and vomiting. And was treated firstly by sigmoid colostomy then pull through operation when she was come back on 25 years old.

CASE REPORT
20 years old female presented to the emergency room with the abdominal pain, vomiting, abdominal distension and absolute constipation for last 4 days.

There is past history of recurrent constipation since birth when her mother noticed that there is delayed passage of meconium till day 4, and then she took her to the medical assistant who did a soap enema and the patient pass gush of meconium and relieved.

From that time the patient had recurrent similar condition and she had previously been hospitalized and treated in such away, but this time the patient condition worse she was toxic, ill, pale ,not jaundice and dehydrated, Bp= 100/60, RR= 30, PR =106.

Upon Abdominal examination she was found to have hugely distented abdomen especially in left
lower quadrant, umbilical is central and everted, no surgical scar or dilated vein, there was tenderness overall abdomen with loud bowel sound and hyper resonant on percussion.

Upon digital rectal examination there was fecal impaction which is very hard on the rectum. All basic and routine investigation were requested CBC: TWBC =12,000 (mainly neutrophil), Platelets =180, Hb=8gm/dl. 

RFT: urea=60 Mlmol/dl, creatinine =1, 7, Na⁺ = 135, K⁺ = 2.1.

We start with resuscitation and rehydration then we put her on: IV fluid, antibiotics, NG tube and regular suction with K⁺ correction and regular enema.

After stabilization patient condition became well we plan for rectal biopsy depending on her history and we did it by proctoscopy and histopathology that revealed there was absence of ganglion cells in myentric (Auerbach) and sum-mucosal (Meissner) plexus in the distal colon which is typical to the classical form of hirschsprung disease. After that we did sigmoid colostomy after bowel preparation and we found hugely dilated sigmoid during the operation. Then we lost patient follow up till she was came back by herself after complete her university and we did pull through operation at age 25 years and now the patient is fine and has no any complications.

**DISCUSSION**

The first documented case of adult HD was described by Rosin et al. in 1950[5]. Nearly 300 cases with a some feature of adult or adolescent HD have been documented. Many more have been documented with a male preponderance, though few studies reported mainly female cases [6, 7].

However in our study the case was female. The term ‘adult HD’ has been arbitrarily applied by some investigators to case in which the patient is older than 10 years when diagnosis is established [8, 9]. Whereas others have defined adult HD as cases in which the diagnosis was made after 18 or 19 years [10]. In Africa, about 20-40% was diagnosed in the neonatal period compared to more than 90% in the western world [11].

With the rarity of Hirschsprung’s disease in adults and the difficulty in diagnosis even in the western world, this explains why only few cases have been documented in Africa [12-14]. And our case is the first case of adult hirschsprung’s disease that reported in Sudan.

The symptoms of adult HD are usually recurrent constipation with abdominal pain and in the clinical examination there’s usually abdominal distention with tenderness which can be associated with palpable fecal mass. And these symptoms usually relieved by enema.

We can use Barium enema or CT scan as imaging tools but in our case we were depend on the patient’s history and the rectal biopsy which is the gold standard tool for the diagnosis because it show the aganglionosis in the sub-mucosal and myentric plexuses.

Preferred treatment with least complications and good quality of life has been Duhamel’s endorectal pull-through [15-17].

In this case presented in this study we treat her by colostomy first and then we make a pull through operation that without complication postoperatively, some cases can present with intestinal obstruction so labrotomy should be considered here.

**CONCLUSION**

So by conclusion HD should be considered in differential diagnosis of any case that presented with recurrent constipation associated abdominal distention and confirmed by aganglionosis in the rectal biopsy which must be treated with pull through operation also must be there’s awareness about atypical presentation as intestinal obstruction that should be treated probably to prevent the complication.

**REFERENCES**