Abstract: Juvenile polyposis syndrome (JPS) is characterised by predisposition to hamartomatous polyps in the gastrointestinal tract, specifically in the stomach, small intestine, colon and rectum. The term juvenile refers to the type of polyp rather than the age of onset. Most individuals with Juvenile polyposis syndrome have some polyps by the age of 20 years, some may have only 4 or 5 polyps over their life time where as others in the same family may have more than hundred. If polyps are left untreated it may cause anemia, bleeding and complications like intussusception. Most juvenile polyps are benign, however malignant transformation can occur. Risk of gastrointestinal cancers ranges from 9 to 50 percentage. Most of the increased risk is attributed to colon cancers. Here I am presenting an interesting case of Juvenile polyposis syndrome who presented with clinical features of acute intestinal obstruction and on laparotomy found to be a case of jejuno-jejunal intussusception.

Keyword: Juvenile polyposis syndrome, jejuno-jejunal intussusception

CASE REPORT:
A 17 year old girl presented in the emergency department with complaints of abdominal pain, vomiting, abdominal distention and obstipation for two days duration. Abdominal pain was of colicky in nature and was relieved by vomiting. Vomiting was of bilious in nature and she had several episodes of vomiting for past 2 days. Along with the above symptoms she also developed abdominal distention and obstipation. She was an already diagnosed case of multiple gastrointestinal polyps. One year back, she presented with complaints of bleeding per rectum. In the process of evaluation she had undergone upper gastrointestinal endoscopy and colonoscopy. She was diagnosed as having multiple gastrointestinal polyps in stomach, duodenum, colon, sigmoid and rectum. Biopsy of polyps were taken during colonoscopy and sent for...
histopathological examination. Histopathology report came as juvenile polyps. No history of any other comorbid illness. No history of similar illness in the family. On examination patient was conscious, oriented, afebrile, dehydrated. Tachycardia present. Per abdomen examination revealed distention and diffuse tenderness. No organomegaly. No gauaring or rigidity. No mass palpable. Bowel sounds - tingling. Aneum examination normal. Rectal examination showed no abnormality. Plane abdominal radiograph revealed multiplet fluid levels and dilated small bowel loops. As clinical features and radiological features were suggestive of acute intestinal obstruction it was thought to be caused by intussusception. Reduction done, but part of jejunum was found to be gangrenous. Gangrenous part of jejunum was resected and jejunojejunal end to end anastomosis. POST OPERATIVE PERIOD-Post operative period was unventful. Oral feeds started on 5th post operative day. Sutures removed on 10th post operative day. Patient discharged on 12th post operative day. Came for review after 2 weeks- no complaints. DISCUSSION-The term polyp is a clinical description of any elevated tumor. Polyps can occur singly, synchronously in small numbers or as part of polyposis syndrome. Juvenile polyposis syndrome is clinically diagnosed if any three of the following present. 1- More than five juvenile polyps of the colon or rectum 2- Juvenile polyps in other parts of the gastrointestinal tract 3- Any number of juvenile polyps and one or more affected family members. Incidence of Juvenile polyposis syndrome is 1 in 100000. Mutation related to Juvenile polyposis syndrome are BMPR1A and SMAD4. Inheritance is Autosomal Dominant. It can run in families or occur sporadically. Associated congenital extracolonic anomalies are described in as many as 20% of these patients, ranging from neurologic (macrocephaly), thoracic (congenital heart disease) and urogenital to gastrointestinal tract (malrotation). Congenital anomalies are seen more common in sporadic cases. Three types of juvenile polyposis syndrome have been described a, Juvenile polyposis of infancy - Most severest form with poorest outcome. Presents with protein losing enteropathy, diarrhoea, anemia, failure to thrive b, Generalised juvenile polyposis- Polyps present through out gastrointestinal tract c, Juvenile polyposis coli- Polyps develop only in colon. The polyps vary in size and number, size and shape. Bleeding and anemia is the commonest presentation. Polyps will predispose to intussusception. Approximately 75% of individual with Juvenile polyposis syndrome have an affected parent. Evaluation includes complete blood count, upper gastrointestinal endoscopy, colonoscopy if patients presents with history of abdominal pain, rectal bleeding, constipation, diarrhoea, change in stool size and shape. Surveillance and genetic testing among family members should be done once diagnosis of polyposis syndrome is made. The most effective management is routine colonoscopy and polypectomy. In some cases removal of all parts of stomach and colon may be necessary. Diagnosed cases and patients who had undergone surgery needs regular follow up with colonoscopy. In established cases of Juvenile polyposis syndrome in families, annual screening is needed. In case of negative screening colonoscopy should be done once in 3 years. CONCLUSION-Intussusception is a known cause for acute intestinal
obstruction. Intestinal polyps are a predisposing factor for intussusception. Above presented case is a diagnosed case of juvenile polyposis syndrome and intussusception due to polyp is rarely seen.

REFERENCES
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