Juvenile Polyposis Syndrome

Juvenile polyposis syndrome (JPS) is a disease that demonstrates an autosomal dominant inheritance pattern and is usually seen in children who are less than five years old of age. This disease is characterized by the growth of multiple hamartomatous polyps throughout the gastrointestinal tract, which includes the stomach, small intestine and colon. Some of the clinical findings include GI distress and anemia since hamartomas are highly vascular. Remember that this disease portends an increased risk for developing colorectal carcinoma.



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Characteristics

Autosomal Dominant
Domino
Juvenile polyposis syndrome demonstrates an autosomal dominant inheritance pattern, meaning that on average 50% of children of an affected parent will have the disease passed on to them.

Children < 5 Years Old
Child with Less Than (5) Hand
Juvenile polyposis syndrome typically affects children who are less than five years of age.

Clinical Findings

Hamartomatous Polyps in Gastrointestinal Tract
Hammerhead Polyps with GI-guy
Juvenile polyposis syndrome is characterized by the growth of multiple hamartomatous polyps throughout the gastrointestinal tract including the stomach, small intestine, and large intestine. Hamartomatous polyps (aka "hamartomas") are benign masses that consist of an excess accumulation of arborizing normal tissue such as mucosal glands, smooth muscle and connective tissue.

GI Distress
GI-guy with Flare-gun
Gastrointestinal distress is a common feature in patients with JPS. This can include abdominal pain, bloating, nausea, constipation, or diarrhea.

Anemia
Anemone
Since the polyps are very friable, they may bleed easily into the lumen of the GI tract. Significant blood loss may cause clinical anemia.

Considerations

Increased Risk of Colorectal Carcinoma
Up-arrow-risk on Colon-hat-wearing Car-gnome
The number of polyps a patient with juvenile polyposis syndrome has during their lifetime ranges from around 5 to more than 100. The larger the number of polyps, the greater the risk of developing colorectal carcinoma. Other gastrointestinal cancers are also possible.

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