Sinistral portal hypertension is a rare condition, which produce isolated gastric varices and upper gastrointestinal bleeding. It is due to splenic vein thrombosis which is often secondary to pancreatic disease (inflammations, tumors). Initial treatment is conservative. After successful conservative treatment early surgery should be considered. UGI Scopy, liver function tests, USG abdomen (with Doppler) and CECT or CT Angiogram are the investigations necessary for the diagnosis. Splenectomy absolutely eliminates the risk from gastric rebleeding. Prognosis depends on the etiology of pancreatic disease. We present a case of 37 year old man who was successfully treated with splenectomy for gastric bleeding caused by Sinistral portal hypertension followed by a literature review of the reported causes, pathophysiology and management of Sinistral portal hypertension.

**Keyword**: Haematemesis, gastric varices, splenic vein thrombosis, splenectomy, pancreatitis

**INTRODUCTION:**
Sinistral portal hypertension is a clinical syndrome of isolated gastric varices in the setting of splenic vein thrombosis, often due to a primary pancreatic pathology with preserved liver function and a patent extrahepatic portal vein.

Sinistral portal hypertension is a rare cause of upper gastrointestinal haemorrhage but it is important to differentiate it from generalized portal hypertension. Due to increased awareness about this condition and advances in diagnostic approaches, patients with sinistral portal hypertension are being identified over the past decades. As most patients of sinistral portal hypertension are asymptomatic or even experience no complications, the exact incidence is still unknown.

**CASE REPORT:**
A 37 year old man with past medical history of alcoholic pancreatitis presented with 2 episodes of hæmatemesis, with about 50 to 60 ml each time and fever which subsided with medicines present for one day. Patient had malena for 2 days. Patient had past history of repeated admissions for alcoholic pancreatitis. Patient was diagnosed to have diabetes 4 years back and on irregular treatment. Patient had jaundice earlier which was treated conservatively.

At admission patient was anaemic and vitals were within normal limits. Abdomen was soft and mild hepatosplenomegaly was present. All the blood and urine investigations including liver function tests were within normal limits except blood sugar. Ultrasonogram abdomen showed hepatomegaly with fatty infiltration and Contrast Enhanced Computerised Tomogram showed chronic calcific pancreatitis with splenic vein thrombosis and splenogastric collaterals. UGI scopy showed fundal varices and portal gastropathy in the fundus and body of stomach with red signs.
Patient was diagnosed as having Sinistral portal hypertension and underwent splenectomy. Spleen was visualized after freeing omental adhesion and the adhesions to its adjacent structures and so the upper midline incision was converted into T shaped incision. There was extensive perisplenic inflammation.

Postoperative period was uneventful and pneumococcal vaccine was given to the patient. Patient had no hematemesis or melena after splenectomy and UGI scopy done one month after the splenectomy was normal.

DISCUSSION
Greenwald and Wasch first outlined the pathophysiology of Sinistral portal hypertension in 1939. Sinistral portal hypertension is a rare, having incidence of less than 1%, but life threatening cause of upper gastric bleeding. It is commonly caused by an occlusive thrombus in the splenic vein. It could lead to formation of lienoclavo(spleno)-gastric varices in order to decompress increased pressure in the splenic vein. Due to its low incidence, it is likely that most cases of sinistral hypertension are initially misdiagnosed as a generalized portal hypertension. Splenic vein thrombosis occurs in 7-20% of patients having pancreatitis or pancreatic neoplasms; however, bleeding occurs in only approximately 5% of these patients.

The name Sinistral portal hypertension is a misnomer since portal pressure is usually within the normal range in these cases. Other synonymous terminologies referring to Sinistral portal hypertension are left sided portal hypertension, segmental, regional, localized, compartmental, lineal, or splenoportal hypertension. The most common pathologies resulting in splenic vein thrombosis or obstruction include chronic pancreatitis, pancreatic pseudocysts and pancreatic neoplasms.

benign neoplasms (rarely cause) adenocarcinoma and functioning and non-functioning islet-cell (neuroendocrine) tumours.

The splenic vein is susceptible in lesions of the pancreas due to its close anatomical course along the superior pancreatic surface. Inflammatory process in the pancreas cause initial damage to the vascular walls and produce venous spasm, venous stasis and thrombosis. Progressive fibrosis (characteristic of chronic pancreatitis) leads to progressive constriction of the splenomesenteric portal axis.

Splenic vein occlusion results in back pressure which is transmitted through its anastomoses with the short gastric and gastroepiploic veins and subsequently via the coronary vein into the portal system. This results in reversal of blood flow in these veins and engorgement of submucosal veins of gastric fundus or formation of gastric varices. The hypertension is confined to the left side of the portal system and is therefore distinct from the common phenomenon of generalised portal hypertension. The diagnosis of sinistral portal hypertension should be considered in all those patients with upper gastrointestinal bleeding associated with splenomegaly and normal liver function tests.

**Gastric varices**
Gastric varices are classified into 4 types based on their location in the stomach

### Gastro-oesophageal varices (GOV)

**Type 1 gastrooesophageal varices**
These extend to the lesser curvature and most common.

**Type 2 gastrooesophageal varices**
These extend to the greater curvature and are associated with high mortality.

### Isolated gastric varices (IGV)
Type 1 isolated gastric varices Those involve only fundal varices and have a high incidence of bleeding. Type 2 isolated gastric varices are gastric varices Those arise at sites other than the fundus, includes varices in the antrum, corpus, pylorus which also the duodenal varices.

**Portal hypertensive gastropathy**
Portal hypertensive gastropathy is the appearance of gastric mucosa seen in patients with portal hypertension. It is characterized by a mosaic like pattern with or without red spots. Mosaic pattern appears as a white reticular network, separating areas of raised red or pink mucosa resembling the skin of snake. When severe, discrete cherry-red spots, pink speckling or scarlatina type rash collectively called as red marks is seen. Characteristic histological finding of portal hypertensive gastropathy is dilated capillaries and venules in the mucosa and submucosa without erosion, inflammation or fibrinous thrombi.
Classification of portal hypertensive gastropathy by Tanoue et al.

Grade I
- Mild reddening

Congestive mucosa Grade II

Severe redness and a fine reticular pattern separating the areas of raised edematous mucosa Grade III

Point bleeding + grade II

Diagnosis
If isolated gastric varices are seen on endoscopy, the diagnosis of sinistral portal hypertension should be considered especially in patients associated with splenomegaly and normal liver function tests.

Advanced diagnostic studies should be performed to clarify the diagnosis. Diagnosis of sinistral portal hypertension is achieved by a combination of:
- Gastroscopy,
- Liver function tests,
- Ultrasound examination (with Doppler) and/or
- Contrast-enhanced CT scan of the abdomen
- Visceral angiography
- CT angiography

Treatment
An expectant management is justifiable in asymptomatic patients with pancreatitis. Symptomatic (Gastric bleed) sinistral portal hypertension is best treated by splenectomy. Patients with chronic pancreatitis undergoing operative treatment should be strongly considered for splenectomy when they have splenic vein thrombosis and associated gastric varices. Less invasive endovascular treatments have shown benefit in selective cases which include splenic artery embolization and transhepatic splenic vein stent placement. Splenic artery embolization should be done in actively bleeding patients who are not medically fit for splenectomy. Splenectomy decreases the inflow into the left portal system by ligation of the splenic artery. It results in decompression of the gastric varices. Prophylactic splenectomy is not necessary in all patients with sinistral portal hypertension. The benefits of splenectomy are obvious in the management of those with severe upper gastrointestinal haemorrhage due to Sinistral portal hypertension in order to rapidly reverse the cause. The overall prognosis for patients with sinistral portal hypertension is clearly dependant on the primary pathology but will invariably be poor in cases with a malignant primary pathology, as involvement of the splenic vein implies advanced infiltrative disease.

CONCLUSION
Sinistral portal hypertension is uncommon cause of haematemesis. Bleeding occurs from isolated gastric varices. Pancreatic disease is the most common etiology. Sinistral portal hypertension should be considered in the presence of gastrointestinal bleeding with normal liver function and unexplained splenomegaly. While splenectomy is the treatment of choice for cases complicated by variceal bleeding, there is no consensus on the treatment of asymptomatic patients. The prognosis of left-sided portal hypertension mainly depends on the underlying etiology.

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