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EHPVO in Children

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Abstract
Background: *EHPVO is an important problem in children. To find out incidence / investigational profile, clinical and therapeutic outcome of children with EHPVO with following study was performed.*
Method: *The children (<12 years) who were admitted in Pediatric GE ward of PGIMER Chandigarh with history of UGI bleed suggestive of EHPVO from July 1993 to June 2003 were noted. Age / sex / clinical details like Haematemesis / Melaena / Splenomegaly / feature of liver disorder (ascites / jaundice / HE etc) / growth parameters / Neonatal history of umbilical catheterization / sepsis / investigation like USG whole abdomen / LFT / PT / Hemogram / UGI Endoscopy etc were noted.*
Results: *Out of 50 cases of EHPVO, 30 were male and 20 female. Age was 3 – 12 years (mean 6 years). History of umbilical catherisation / sepsis were present in 6% cases. Fifty out of 50 (100%) came with Haematemesis / 30 (60%) with combined Haematemesis / Melaena. There was no feature of liver failure in any case. USG shows normal hepatic echo texture with 40 cases (80%) having portal cavernoma, 30 (60%) having both portal cavernoma and splenic collateral, 10 (20%) having peri cholecystic collateral. Splenomegaly was present in all cases (100%). There was no colorectal Varix or Hemorrhoid. Gastic varix develop only after EVL / EST of Esophageal varix in 10 (20%) cases. EVL was successfully done 40 (80%) cases but 10 cases (20%) required EST due to technical problem. 5 (10%) cases required distal splenorenal shunt. All patients of EHPVO recovered from portal hypertension but 10 (20%) had growth failure.*
Conclusion: *EHPVO being important cause of portal hypertension requires urgent management.*

Introduction

Extra Hepatic portal venous obstruction (EHPVO) is commonest cause of portal hypertension and vericeal bleed in children. It is characterized by massive slpenomegaly hypersplenism, growth failure, ectopic varix (eg: rectal varix, portal biliopathy etc). Procoagulant state is not commonly seen in children unlike adults. Growth failure is due to reduced liver blood flow, insulin delivery to liver and low IGF-1[(1)]

In our series, we have not come across colorectal varix / hemorrhoid.

In EHPVO portal vein is converted to portal cavernoma at portahepatis – resulting in GE varies / portal hypertension. Pericholecystic, paracholedocholic, para pancreatic, duodenal vein, collateral are seen. Portal biliopathy, hypersplenism, growth retardation are seen. Endoscopic band ligation / sclera therapy / porto systemic shunt surgery are done. Through liver

function is normal but functional improvement can happen in long term.[2]

EHPVO and cirrhosis are two major causes of portal hypertension in children. But predominant cause of vericeal bleed is EHPVO.[3]

Endoscopic band ligation (EVL) is much better than endoscopic sclero therapy (EST) alone. Because EVL plus EST have fewer complication than EST alone.[4]

EVL and EST are directly compared to each other in some study. It is found that EVL is safe and more effective in eradicating varices than EST. EVL has less number of re-bleed / fewer complication / quicker eradication of varices compared to EST.[5]

EVL in children with EHPVO is done successfully but there is significant increase in IGV (Isolated Gastric Varix), significant decrease in GOV1 (Gastric Esophageal Varix No-1), increased frequency of PHG (Portal Hypertensive Gastropathy.[6]

In our series gastric varix developed in 10 (20%) cases of the EVL/EST.

EST for esophageal varix in children with EHPVO is safe in follow up study as there is no significant re-bleed in follow up.[7]

15 year follow up of EST in children showed that EST safe and effective in essophageal variceal bleed. It prevents 88% bleeding after vericeal eradication. Surgery is done as a complimentary technique in EHPVO where there is significant gastric bleed / painful sclero therapy / GAVE (Gastric Antral and Variceal Ectasia Formation). [8]

PHG and Gastric varices are common in children with EHPVO following EST. There is clear evidence of varices development.[9]

Non cirrhotic portal hypertension (NCPH) and EHPVO are two prototype of this category where portal hypertension occurs without cirrhosis. Infective / prothrombotic state – are two etiological factors for NCPH.

Variceal bleed / massive splenomegaly / portal biliopathy / parenchymal extinction after prolonged portal hypertension makes less favourable prognosis for NCPH. Nodular regenerative hyperplasia is a distinctive feature of NCPH.[10]

EHPVO is disease of children but NCPH is disease of young adult / middle aged women.

In both cases, heterogeous group of liver disorder are found. They are mostly of vascular origin – leading to portal hypertension / near normal HVPG (Hepatic Venous Pressure Gradient). In EHPVO, early age acute or recurrent infection with thrombotic origin predisposes to portal hypertension. But umbilical sepsis is found in only 6% of cases.

In EHPVO, liver function is preserved but growth failure / portal biliopathy are important feature.

Shunt surgery is done if endoscopic procedure fails / symptomatic hypersplenism / biliopathy / ectopic varix / remote residence of patients where endoscopic therapy is unavailable.[11]

In non cirrhotic portal fibrosis (NCPF) there is intrahepatic / prehepatic lesion. Lesion is of vascular origin – portal vein / its branches / perisinusoidal area. HVPG is normal.

NCPF has obscure origin. Low socio economic condition / well tolerated variceal bleed / obstructive portal venopathy / mean normal liver function / marginal splenomegaly are characteristic of NCPF.

NCPF is often called idiopathic portal hypertension (IPH) or hepato portal salerosis.

EST / EVL can eradicate varix in 85 – 90% cases. Gastric varix is tackled by cyano acralitic glue or surgery. surgery is indicated when endotherapy fails or symptomatic hypersplenism.[12]

Portal hypertension without cirrhosis is called non cirrhotic portal hypertension (NCPH). Causes of NCPH are non cirrhotic portal hypertension (NCPF) / extra hepatic portal venous obstruction (EHPVO).

Other causes of NCPH are hepatic schistosomiasis, hepatic vein out flow obstruction, veno occlusive disease, congenital hepatic fibrosis. Diagnosis is done by USG / UGI endoscopy / normal LFT / normal liver biopsy. Morbidity and mortality from variceal bleed is

lesser than cirrhosis because liver function is normal.

Treatment of NCPF is EST / EVL / Beta blocker / splenectomy / porto systemic shunt.[13]

Methods

The children (<12 years) who were admitted in Pediatric GE ward of PGIMER Chandigarh with history of UGI bleed suggestive of EHPVO from July 1993 to June 2003 were noted. Age / sex / clinical details like Haematemesis / Melaena / Splenomegaly / feature of liver (ascites / jaundice / HE etc) / growth parameters / Neonatal history of umbilical catheterization / sepsis / investigation like USG whole abdomen / LFT / PT / Hemogram / UGI Endoscopy etc where noted.

Results

Out of 50 cases of EHPVO 30 were male and 20 female. Age was 3 – 12 years (mean 6 years). History of umbilical catherisation / sepsis were present in 6% cases. Fifty out of 50 (100%) came with Haematemesis / 30 (60%) with combined Haematemesis / Melaena. There was no feature of liver failure in any case. USG shows normal hepatic echo texture with 40 cases (80%) having portal cavernoma, 30 (60%) having both portal cavernoma and splenic collateral, 10 (20%) having peri cholecystic collateral. Splenomegaly was present in all cases (100%). There was no colorectal Varix or Hemorrhoid. Gastic varix develop only after EVL / EST of Esophageal varix in 10 (20%) cases. EVL was successfully done 40 (80%) cases but 10 cases (20%) required EST due to technical problem. 5 (10%) cases required distal splenorenal shunt. All patients of EHPVO recovered from portal hypertension but 10 (20%) had growth failure.

Discussion

Portal hypertension in children are EHPVO (54%), Cirrhosis (39%), CHF (3%), NCPF (2%), Budd chiari syndrome (2%).

UGI bleed from portal hypertension in children are EHPVO (85%), cirrhosis (10%), CHF (2.8%), NCPF (2%), Budd chiari syndrome (1%).

Mortality from bleed is 1.7% in EHPVO but 30% in cirrhosis.[3]

Cavernoma transformation around portal vein is seen in EHPVO. Extra hepatic collateral channels are seen, splenic collaterals, porto systemic collaterals are seen around Lt gastric vein, peri splenic vein, peri cholecystic collaterals are also seen.[14]

Surgical treatment of EHPVO has dramatically improved nowadays.

In our series 5 (10%) cases required distal splenorenal shunt.

Meso Rex bypass shunt is used widely but limited by favourable anatomy.[15]

Mesenteric Lt portal vein bypass in children with EHPVO has become popular.

Mesenteric portal Rex shunt is therapy of choice for EHPVO. It abolishes hepato pulmonary syndrome.[16]

Mesenteric Lt portal bypass (Rex shunt) is preferred to other surgical procedure because it restores normal portal flow to liver. It eliminates portal hypertension and its consequences but restores normal liver function.[17]

Both meso Rex shunt and porto systemic shunt relieves symptoms of portal hypertensive bleeding in children. But Meso Rex better relieves hypersplenism. It restores normal portal venous pressure and maintain normal flow to liver.[18]

Children of EHPVO suffer from malnutrition and growth hormone resistance. It leads to growth retardation. IGF – I increases after Meso Rex shunt.[19]

In Meso Rex by pass – autologous Lt internal jugular vein graft is used to bypass portal blood circulation. Other vascular conduits are autologous saphenus vein, splenic vein, Rt gastro epiploic vein, inferior mesenteric vein.

Umbilical veins harvested from deceased liver are processed – dilated – channelized for patency – connected with Lt portal vein.

Cross section of recanalised umbilical vein undergoes histological examination / stained with heamatoxilin eosin / immunohisto chemistry for CD 31 / Factor VIII antigen.

Modified Meso Rex shunt is done by umbilical vein.[20]

Risk factor for EHPVO is detected in 43% cases. 30% has perinatal origin. 13% has prothrombotic state.

Prothrombotic state is mainly due to protein c / protein S deficiency, lupus anti coagulant etc.

Splenomegaly is seen in 43% cases and UGI bleed in 40% cases.[21]

Colorectal variceal bleed is 0.5% of all EHPVO bleeding cases. Colonic varix is detected by colonoscopy / angiography. Transanal ligature of hemorrhoids / rectal varix is done. Surgical procedure may also be necessary.[22]

In follow up study of 40 yrs, porto systemic shunt is proved to be effective therapy for bleeding varix in EHPVO.

In many years of surveillance, freedom from recurrent bleed / normal LFT / no encephalopathy is noted.[23]

EVL plus EST eradicates varix in larger number of patients than EVL alone with no extra complications.[24]

Long term follow up of EHPVO is done for hypersplenism / porto biliopathy / variceal bleed / neuropsychiatric disease / growth failure.

In our series, an EHPVO children recovered from portal hypertension, but 10 (29%) had growth failure.

Meso Rex shunt is recommended where it is feasible.[25]

Hepatic dysfunction (i.e. Ascites / deranged LFT) is not uncommon in EHPVO with prolonged portal hypertension. EHPVO can lead to progressive liver failure.[26]

75% of EHPVO with minimal hepatic encephalopathy (HE) continue to have minimal HE. New onset minimal HE develop in 5% over 14 Yrs.

EHPVO with minimal HE does not progress to overt HE.[27]

Association of umbilical vein catheterization and sepsis with portal vein thrombosis is inconclusive.[28]

Meso Portal bypass (MPB) for EHPVO in children is treatment of choice where it is technically feasible. MPB provides long term correction of portal hypertension in EHPVO. Left internal jugular vein / recanalized umbilical vein / gastric vein / large colic vein are utilized for MPB graft.[29]

Home delivery / umbilical sepsis are risk factors in development of EHPVO EST can eradicate varix.[30]

Presence of UGI bleed but absence of jaundice is 97% accurate in diagnosis of EHPVO.[31]

In a study of 15 yrs follow up after porto systemic shunt – one had post systemic encephalopathy. One had shunt stenosis requiring angioplasty. Distal splenorenal shunt has excellent prognosis.[32]

Mesenteric Lt. portal bypass (MLPB) functions well in patient with portal hypertension caused by portal vein thrombosis. Physiologic advantage of MPLB are that it reduces portal pressure without reducing hepatic vascular flow.[33]

Mesenteric Lt. portal vein bypass (Rex Shunt) is treatment of choice in EHPVO with portal hypertension provided it is technically feasible. It reduces hepato pulmonary syndrome.[34]

Although idiopathic in etiologies, genetic and acquired thrombophilia is implicated in EHPVO. Post operative Meso Rex bypass thrombosis is corrected by perioperative anticoagnlant strategies for pre operative thrombophilia.[35]

EHPVO has features of hyperdynamic circulation with increased cardiac index / low systemic and pulmonary vascular resistance index (PVRI). These changes are similar to that of cirrhosis. Portal hypertension per se is responsible for genesis of systemic / pulmonary hemodynamic changes.[36]

93% of portal hypertension in children is due to intrahepatic cause and 7% is due to EHPVO.[37]

Management of EHPVO portal hypertension in children include EST / EVL / Vasopressin / SB

Tube / Surgery (TIPSS / Porto systemic shunt) and shunt surgery (Meso caval / porto caval / distal spleno renal / Meso Rex bypass). Placement of autologous venous graft bridges between mesenteric vein to intrahepatic Lt. portal vein. Lt. renal vein and splenic vein are connected by selective shunt (distal) or non selective (central) shunt. A graft between mesenteric or portal vein to IVC can decompress portal vein but graft thrombosis can happen. Even retrograde flow can occur. Porto caval shunt is excellent for lowering portal pressure but chance of hepatic encephalopathy still remains.[38]

Combined EVL plus EST can eradicate in a significantly huge number of patients than EV alone with no extra complication.[39]

Portal hypertension in children are largely due to NCPF (48%) / EHPVO (36%) / Cirrhosis (16%).[40]

Conclusion

EHPVO being an important cause of portal hypertension in children requires urgent management / meticulous follow up for eradication of varix. High index of suspicion is important for recognition of this illness.

Conflict of interest – Nil

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