Extra Hepatic Portal Vein Obstruction – Unobstructed

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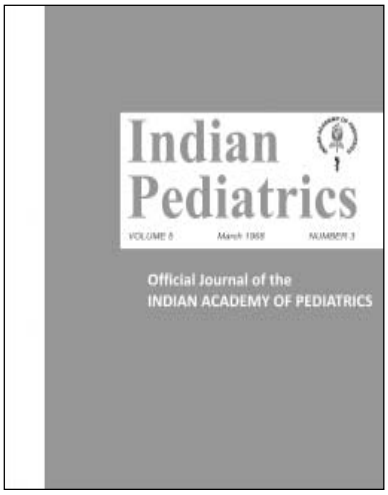
In the March 1968 issue of *Indian Pediatrics*, the published research articles pertained to nephrotic syndrome (clinico-pathological profile), extrahepatic portal obstruction (EHPO), chromosomal analyses of children with mental and physical defects, infectious-allergic myocarditis and megaloblastic anemia. In this write-up, we showcase what was being done in the 1960's in management of EHPO in contrast to what is known and being practised now.



THE PAST

The study [1] entitled 'Extrahepatic portal obstruction in children' was a case series from Medical College, Nagpur, and had the objectives of studying the clinical profile of children with EHPO in terms of presentation, management and outcome. The series comprised of 12 cases; ages ranging from 9 months to 13 years. The investigations performed in all children included hemogram, liver function test, liver biopsy and percutaneous transplenic portal venography. Esophageal radiographs, barium swallow and intra-splenic measurement were performed in a few.

The presenting complaint was upper gastrointestinal (UGI) bleeding in 9 patients and unexplained splenomegaly in 3 patients. The salient clinical signs observed were splenomegaly in all, mild hepatomegaly in 6, and ascites in 2 patients. None had any biochemical evidence of liver dysfunction. Venography revealed extrahepatic block (at hepatic hilum in 9 and at distal splenic vein in 3 patients), vascular collaterals and gastro-esophageal varices in all patients. Initially, all patients were managed conservatively; however, recurrent hematemesis occurred in five patients. One died and 4 underwent surgery that comprised of splenorenal shunt with splenectomy in two, and isolated splenectomy in two patients. The authors concluded that a diagnosis of EHPO could be made clinically based on hematemesis, fluctuating splenomegaly (regression of spleen on acute bleed) and the absence of evidence of hepatocellular failure. However, a splenoportal venogram and liver biopsy were essential for confirmation and planning treatment. They advocated conservative management initially, with surgery reserved only for recurrent UGI bleeding.

Historical background and past knowledge: In 1868, Balfour, *et al.* first described the cavernomatous transformation of the portal vein (PV). Portal hypertension (PHT) was a known clinical entity by the turn of the century and in 1906, Gilbert, *et al.* noted that it was the cause of many cases of UGI haemorrhage with splenomegaly. In 1955, Gibson, *et al.* [2] concluded that the cavernomatous transformation of the PV was secondary to PV thrombosis, rather than a congenital anomaly. By this time, Indian researchers had also joined the bandwagon, describing the trans-splenic portal venography patterns in EHPO. In 1962, Shaldon and Sherlock [3], on the basis of clinical profile of 16 cases, concluded that extrahepatic PHT was a syndrome of splenomegaly, gastrointestinal hemorrhage, anemia and portal thrombosis in the absence of cirrhosis. It was recognized that it differed from hepatic PHT in terms of clinical profile, management and outcomes, and that an association with umbilical sepsis existed.

THE PRESENT

Over the years, nomenclature has changed to extra hepatic portal vein obstruction (EHPVO), with the addition of 'vein' to the pre-existing term. It is reportedly the commonest cause of portal hypertension in Indian children, unlike in Western countries where hepatic dysfunction is more common [4]. Though it is widely postulated that umbilical sepsis, umbilical vein catheterization, abdominal trauma, surgery, intra-abdominal sepsis, dehydration, congenital agenesis and portal vein atresia may result in EHPVO, there is still

paucity of strong supportive scientific evidence to substantiate this. Deficiencies of antithrombotic factors (protein-C, protein-S, anti-thrombin III) have frequently been found in EHPVO. These have been explained by low synthesis due to the poor hepatic blood flow resulting from the PV thrombosis and increased clearance due to the portosystemic shunt [5].

In the late 1970's and 1980's, ultrasonography (USG) replaced visceral angiography for evaluating portal venous system and porta-systemic anastomoses in most children. Computed tomographic (CT) portal venography and magnetic resonance imaging (MRI) emerged in the 1990's as effective and non-invasive tools to visualize the portal venous system in pre- and post-shunt cases. In the late 1990's, ultrasound Doppler superseded conventional USG as a more sensitive method, and remains the first modality of choice till date. When inconclusive, a CT portal venography or MRI may be required for demonstrating portal vein obstruction.

Management includes treatment of variceal bleeding, and co-morbidities like growth failure, portal biliopathy, hypersplenism and physical disability. In acute variceal bleeding, hemodynamic resuscitation should be followed by either endoscopic sclerotherapy (EST) or endoscopic variceal band ligation (EVL) or both. These modalities can also prevent further bleeding. The eradication rate of esophageal varices is 88-100% with EST, but complications like ulcers and strictures are common. A randomized control trial that compared EVL *versus* EST in 49 children with EHPO showed that EVL achieved variceal eradication more quickly, required fewer sessions, had lower re-bleeding rate and fewer complications compared with sclerotherapy; although both were equally effective in arresting acute bleed and had similar recurrence rate [6].

Conventional shunt (proximal or distal spleno-renal) surgery is indicated in situations when there is failure of endoscopic therapy, when gastric or ectopic varices are not amenable to endoscopy, symptomatic hypersplenism, growth retardation, portal biliopathy, massive splenomegaly affecting quality of life, or when arranging immediate blood transfusion is difficult. Presently, endoscopic therapy is a lifesaving modality in reducing acute variceal bleed related mortality. It may remain the sole therapy for most patients and serve as a bridge therapy in those where definitive surgery is warranted.

A major breakthrough that has revolutionized surgical management in the last decade has been the introduction of the mesenterico-portal bypass (MPB) or Rex shunt. The internal jugular vein is used as a conduit to bypass blood from the superior mesenteric vein to the left branch of the PV, thus closely simulating a physiological state. This restores hepatic blood flow, corrects PHT and improves systemic manifestations of EHPVO and linear growth [7]. It is now emerging as the optimal mode of therapy. The paradigm shift in practice that has emerged is surgery as soon as the diagnosis is established, rather than later as an alternative resort. This is because the ability of the portal venous system to adapt to restored flow is age-dependent and prolonged deprivation of blood flow may lead to atrophy of intrahepatic portal veins [8]. However, unfavourable anatomy in a large number of cases, requirement of an interventional radiologist, and paucity of surgical experience have limited its application in most settings at present.

There has been a huge leap forward in the management of EHPO in the last 50 years, from invasive to non-invasive diagnostic modalities to the recommendations of early shunt surgery. These have paved the way for improvement in quality of life and prolonged life expectancy in these patients. With the availability of greater resources, wider accessibility and better surgical expertise, the future certainly appears promising.

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