

Portal Vein Thrombosis in Sudanese Children

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Abstract:

Background: Portal Vein Thrombosis (PVT) is one of the most common causes of portal hypertension among children in Sudan.

Objectives: To determine the incidence, aetiology and mode of presentation of PVT among children and find its relation to gastrointestinal bleeding in a Sudanese hospital.

Methodology: This is a prospective hospital based study conducted at a tertiary care paediatric hospital in Khartoum, Sudan (Gaffer Ibn Oaf Specialized Children Hospital (GIO)). 350 children were diagnosed on clinical grounds (heamatemiss, splenomegaly) to have portal hypertension. After laboratory investigations (CBC, LFTs, coagulations profile and RFTs), abdominal sonography with Doppler, 275 patients turned to have portal vein thrombosis. Upper GI endoscopy was done for all patients and liver biopsy when indicated. Data were analysed, discussed and conclusion were reached.

Results: Out of 800 children presented with upper GI bleeding 350 were diagnosed as having esophageal varices bleeding due to portal hypertension (44%), out of these 250 (70%) children were found to have PVT as a cause of their portal hypertension, and another 25 children were diagnosed to have PVT without GI bleeding. Their ages ranged between 4 month and 16 years. Two hundred were males. Seventy five were below one year of age. History of risk factor for PVT was found only in 68 children. All the bleeders had sclerotherapy, band ligation or both beside propranolol and H₂ blockers or PPI. Time for follow up was between 6 month and 4.5 years. 150 children cleared their varices with 4-6 session of either sclerotherapy, banding or both, the rest of the children are still receiving sclerotherapy. Ten children received liver support for portal biliopathy, five children had splenectomy because of hypersplenism. Four children died. Propranolol was stopped in 50 children out of 150 children who cleared their varices after two years

Conclusions: PVT in children is common in Sudanese children, and represents the second common cause of upper GI bleeding. Its aetiology is obscure in the majority of cases and more studies and facilities are needed to uncover the underlying cause. Simple clinical data, ultrasonography and upper GI endoscopy are quite adequate measures to reach accurate diagnosis.

Combinations of endoscopic sclerotherapy and band ligations with propranolol are very effective in controlling the bleeding of the esophageal varices, though it has a remarkable effect on the quality of life of affected children. Thus appropriate medical alternative or surgical treatments are needed in order to reduce morbidity and mortality and improve the quality of life of these patients.

Keywords: Portal hypertension, sclerotherapy, band ligation, biliopathy, splenectomy.

Portal Vein Thrombosis (PVT) is one of the most common causes of Portal Hypertension among children in Sudan¹.

The incidence, aetiology, clinical manifestation, treatment modules and outcome were not studied before in Sudan.

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Objectives

To determine the incidence, aetiology and mode of presentation of PVT among children who presented to our Gastrointestinal (GI) unit at Gaffer Ibn Oaf Specialized Children Hospital (GIO) in Khartoum, during January 2005-June 2010, and see how many of them presented with GI bleeding. Also we want to determine the management and out the outcome of the treatment over that period of time.

Patients and Method: This study was conducted at GIO Children Hospital, Khartoum, Sudan, which is the main tertiary care paediatric hospital and represents the only centre for established paediatric endoscopy unit in the country. Out of 5000 children seen at our GI unit over the last five years, 275 were diagnosed to have PVT. Detailed history was obtained for all the children with special references to the risk factors (umbilical sepsis, cathetrization, and admission to SCABU, gastroenteritis and dehydration or septicaemia) and history of consanguinity were obtained. Their ages ranged between four month and 16 years. The diagnosis was made on clinical interpretation (heamatemiss, splenomegaly, portalbiliopathy), laboratory investigations (CBC, LFTs, coagulations profile and RFTs), abdominal sonography with Doppler, upper GI endoscopy and liver biopsy when indicated.

We couldn't perform the haemophilic screen for technical reasons, we managed only to do it in two children on their family's expenses abroad and it was negative.

Results: Out of 800 children presented with upper GI bleeding 350 were diagnosed as having oesophageal varices bleeding due to portal hypertension (PHT) (44%). Out of these 250 (70%) have PVT as a cause of their PHT. 100(30%) children have other causes.

Another 25 children diagnosed with PVT who did not have GI bleeding, (10 children presented with hyper splenism, 10 with portal biliopathy and five presented with only splenomegaly).

Two hundred children were males. Seventy five were below one year of age and they all presented with heamtemisis with or without splenomegaly.

History of risk factors (umbilical sepsis, cathetrization, and admission to SCABU, gastroenteritis and dehydration or septicaemia) was found in 68 children and consanguinity in 75 children, and both in only 40 children.

Management was focused to treat complications and included primary and secondary prophylaxis against upper

gastrointestinal bleeding (that resulted from the rupture esophageal varices). All the bleeders had sclerotherapy, band ligation or both beside propranolol and H₂ blockers or PPI.

Ten children received liver support for portal biliopathy, five children had splenectomy because of hypersplenism. None of those children had any form of portocaval shunt, nor other surgical interventions.

Time for follow up was between 6 month-4.5 years. 150 children cleared their varices with 4-6 cession of either sclerotherapy, banding or both, the rest of the children are still receiving sclerotherapy. We lost two children in the hospital with severe upper GI bleeding and another two children with bleeding at home.

Propranolol was stopped in 50 children out of 150 children who cleared their varices after two years; they developed good porto-systemic collaterals evident by ultrasonography and Doppler.

Discussion

Similar to reports from elsewhere, the cause of PVT is not known in the majority (73%) of our patients^{2,3}.

Detailed history revealed risk factors for PVT in around 27%, consanguinity in 40% and both in around 15%. Almost all the families couldn't afford to do the haemophilic screen apart from two children in whom the screen was done abroad and it was negative. However, in our patients, the low socioeconomic class as a separate risk factor or its association with umbilical sepsis, gastroenteritis, dehydration and malnutrition might have contributed to that⁴.

The main presenting symptom was upper gastrointestinal bleeding in 91% of the children which is consistent with the literature⁴, while splenomegaly was found in the majority of them, however, pancytopenia due to hyper splenism was found in only five children. Hepatopulmonary syndrome is rare in non-cirrhotic patients who have PVT, in line with that none of our patients had the syndrome⁵.

Abdominal ultrasound was reported to have high sensitivity (94-100%) and specificity

(90-96%) in the diagnosis of PVT, upper GI endoscopy will support that and aid the management especially in bleeders^{2, 5}. In concordance with that abdominal ultrasonography and upper GI endoscopy were able to suggest the diagnosis in almost all of our patients. As expected LFTs were normal in the vast majority apart from only less than 1% in whom liver biopsies were performed and revealed periportal fibrosis and that goes with the literature³.

Sclerotherapy combined with band ligations (when technically possible) were very effective in clearing the bleeding oesophageal varices electively⁶.

Supporting that in 150 children, we managed to eradicate the varices after 4-6 intervention sessions, 6 weeks apart in average, and the patients were continued on prophylactic therapy. The rest are still receiving sclerotherapy sessions and continuing to receive propranolol. Propranolol was very effective as prophylaxis to prevent further re-bleeding⁷. Fifty of the children who cleared the varices needed no more propranolol.

This form of treatment, though safe and effective (with mortality of less than 0.1%)⁸⁻¹¹, affects the children life and family so adversely, with the amount of hospital admissions, the frequent endoscopic sessions and the medications they have to take. Most of the older children had become school leavers.

The outcome of selective portocaval shunts seems to be quite encouraging¹². The applicability of such technique in Sudan is not an option in the present time with the lack of trained paediatric vascular surgeons, and hence none of our patients had this procedure. Splenectomy was necessary in five children because of hypersplenism, though we noticed the degree of splenomegaly spontaneously decreased with time, probably indicating the development of porto systemic collaterals. This is consistent with the findings that childhood PVT bleeding tend to decrease in early adulthood¹³.

Children who had biliopathy are on liver support and conservative support and

management, though they all had their growth being compromised¹⁴.

Mortality rate in non-cirrhotic variceal bleeders is not high³, going with that only four of our patients died because of bleeding. Although it has been postulated that the cause of growth retardation is due to chronic anemia, intestinal venous congestion leading to malabsorption syndrome, hepatic hypoperfusion resulting in hepatotrophic hormone deficiency^{2,15, 16}, the reason why some of the children with PVT have their growth compromised while the others don't seem to be affected remains a challenge that needs to be studied more!!

Conclusions

PVT in children is common in Sudanese children, and represents the second common cause of upper GI bleeding. Its aetiology is obscure in the majority of cases and more studies and facilities are needed to uncover the mysterious about it. Nevertheless, the common known risk factors and thrombophilic seem to contribute to a significant number.

Simple clinical data, ultrasonography and upper GI endoscopy are quite adequate measures to reach accurate diagnosis.

Combinations of endoscopic sclerotherapy and band ligations with propranolol are very effective in controlling the bleeding of the esophageal varices, though it has a remarkable effect on the quality of life of affected children. Thus appropriate alternative surgical treatments are needed in order to reduce morbidity and mortality and improve the quality of life of these patients.

References

1. Omayma MS, MO EH Gadour. Upper Gastrointestinal Endoscopy in Sudanese Infants and Children :SudanJMS,vol.2,no2,June2007.91-94
2. Sarin SK, Agarwal SR. Extrahepatic portal vein obstruction. *SeminLiver Dis.* 2002;22:43-58. .
3. Schettino GCM, Eleonora D T. Fagundes EDT, RoqueteMLV et al. Portal vein thrombosis in children and adolescents *Jornal de Pediatria* 2006;82(3): 171-178.
4. Alvarez F, Bernard O, Brunelle F et al. Portal obstruction in children. Clinical investigation and hemorrhage risk. *J Pediatr.* 1983;103: 696-702.

5. Gürakan F, Eren M, Kocak N et al. Extrahepatic portal vein thrombosis in children: etiology and longterm follow-up. *J Clin Gastroenterol.* 2004;38: 368-72.
6. Balistreri WF. Desenvolvimento e função. In: Behrman RE, Kliegman RM, Jenson HB. *Nelson textbook of pediatrics.* 16th ed. Philadelphia: WB Saunders; 2002. p. 1179-82.
7. Toppa NH, Andrade MC. Princípios gerais da histopatologia hepática. In: Penna FJ, Mota JAC, Roquete MLV, Ottoni CMC, editores. *Doenças do fígado e vias biliares na infância - Parte 1.* Rio de Janeiro: Medsi; 1996. p. 129-48.
8. Gardner E, Gray DJ, O'Rahilly R. *Anatomy - a regional study of human structure.* 4th ed. Philadelphia: WB Saunders; 1975. p. 385-96.
9. Carvalho AST, Mendes CMC. Hipertensão porta: tratamento. In: Penna FJ, Mota JAC, Roquete MLV, Ottoni CMC, editores. *Doenças do fígado e das vias biliares na infância - Parte 2.* Rio de Janeiro: Medsi; 1999. p. 259-94.
10. Pinto RB, Vieira SM, Silveira TR. Hipertensão porta. In: Ferreira CT, Carvalho E, Silva LR. *Gastroenterologia e hepatologia em pediatria. Diagnóstico e tratamento.* Rio de Janeiro: Medsi; 2003. p. 683-707.
11. Silva RAP. Ultra-sonografia das doenças hepáticas e biliares na infância. In: Penna FJ, Mota JAC, Roquete MLV, Ottoni CMC, editores. *Doenças do fígado e vias biliares na infância - Parte 1.* Rio de Janeiro: Medsi; 1996. p. 45-88.
12. Shneider BL. Portal hypertension. In: Suchy FJ, Sokol RJ, Balistreri WF, editors. *Liver disease in children.* 2nd ed. Philadelphia: LWW.com; 2001. p. 129-51.
13. Valla DC, Condat B. Portal vein thrombosis in adults: pathophysiology, pathogenesis and management. *J Hepatol.* 2000;32:865-71.
14. Orloff MJ, Orloff MS, Girard B et al. Bleeding esophagogastric varices from extrahepatic portal hypertension: 40 years' experience with portal-systemic shunt. *J Am Coll Surg.* 2002;194:717-28; discussion 728-30.
15. Kato T, Romero R, Koutouby R et al. Portosystemic shunting in children during the era of endoscopic therapy: improved postoperative growth parameters. *J Pediatr Gastroenterol Nutr.* 2000;30: 419-25.
16. Bucuvalas JC, Cutfield W, Horn J et al. Resistance to the growth-promoting and metabolic effects of growth hormone in children with chronic liver disease. *J Pediatr.* 1990;117:397-402.