Portal Hypertension on Cavernoma in Children: Advanced Surgical Therapeutics (About 10 Cases)
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Abstract: The portal cavernoma is the most common cause of portal hypertension in children responsible for important morbidity linked to gastrointestinal bleeding. This study aims to elucidate the main therapeutic advances in the surgical treatment of portal cavernoma. We are conducting a retrospective study on 10 children affected by portal cavernoma and compiled at surgery "A" of Rabat children's hospital between 2000 and 2017. The average age of patients was 10.7 years and the sex ratio was 1. The reason for hospitalization was gastro-intestinal bleeding in 7 cases and splenomegaly in 3 cases. Biology has shown hypersplenism in all our patients, the prothrombin time (PT) was elongated in 8 cases and liver function tests were abnormal in 1 case. The gastrointestinal endoscopy showed oesophageal varices steadily. The ultrasound, with or without a Doppler, established the diagnosis in all cases. The treatment was endoscopic in 2 cases and surgical in 8 cases consisting of a distal splenorenal shunt in 2 cases, a spleno-caval shunt in 1 case, a meso-caval venous interposition shunt in 2 cases and splenectomy in 3 cases. The evolution of the patients was generally good with permeable shunts and a regression of hypersplenism. Also, no recurrence of bleeding was seen in 8 cases. However, 2 of the surgical interventions were complicated by portal vein thrombosis. This study has enabled us to understand better the pathology of portal cavernoma and its repercussions that could be disastrous. Accordingly, an appropriate therapeutic decision must be taken as soon as possible especially after the advent of Mesoporal bypass.

Keywords: Portal hypertension – portal cavernoma – Rex-shunt – Portosystemic shunt.

INTRODUCTION
A portal cavernoma is a venous vascular anomaly that appears after a chronic occlusion (at least three weeks old), often thrombotic, of the extrahepatic portal system, upstream of the liver. It consists of a network of veins within which travels a hepatopetal portal blood. These veins, initially millimetric or microscopic caliber, dilate and become tortuous forming a veritable network of angiomatos pace which is still unable to ensure the drainage of the entire splanchnic circulation, hence the installation of portal hypertension.

In children, cavernoma is a common cause of extrahepatic portal hypertension called “pre- or infrahepatic” very often manifested by gastrointestinal bleeding, related to variceal bleeding, which can be life-threatening.

The positive diagnosis, previously done intraoperatively, is currently easily used in imaging. However, the etiological diagnosis remains mysterious in the vast majority of cases. Therapeutic management, especially surgery, has also been progressing thanks to the advent of portal reperfusion.

The present work aims to project the light on therapeutic surgical advances based on the experience of the A - HER pediatric surgery department in order to compare our results with those of the literature.

MATERIALS & METHODS
This is a retrospective study that focuses on a series of 10 children admitted for portal hypertension on cavernoma and having undergone surgical or endoscopic treatment during the period 2000-2018. Inpatients with portal hypertension who did not have cavernoma were excluded as the etiological diagnosis of portal hypertension.

The sex ratio is 1 with an attack of five girls against five boys. The average age of intervention is about 10 years. (10.7 years), ranging from 5 to 14 years old. The concept of consanguinity of the parents was found 3 cases. None of our patients have a history of neonatal hospitalization, umbilical catheterization,
abdominal trauma or infection. No family history reported thromboembolic.

2 children had a history of geophagy.

A child has been operated on for scrotal hernia six years before the onset of symptoms (Case 9). Digestive haemorrhages prompted consultation in 7 cases (70%). The SMG was involved in the hospitalization of 3 children (30%), painful in 2 cases (Case 2 and 6) and accidental discovery in 1 case (Case 4). Externalized haemorrhage was present in 7 cases (70%) and absent in only one case (Case 6). It was digestive in 7 cases and extradigestive (epistaxis) in 2 cases. These haemorrhages caused shock in 2 cases (Case 1 and 3). Abdominal pain was recorded in 3 patients (30%). Mucocutaneous pallor was constant in 100% of cases. 2 children had fever (20%). The CVC was scored in 5 cases (50%). A SMG was palpable in 100% of cases. HMG was found in 3 children (30%).

No dullness indicative of ascites was found (0%). Lack of jaundice in children of our series (0%). A blood count was performed in all the patients of our series and affirmed in 100% of the cases the existence of a hypersplenism, with a pancytopenia in 9 cases (90%) and a bicytopenia in 1 case (Case 3).

Hypochromic microcytic anemia was constant in 10 cases and severe in 5 cases (Hb <7) with an average Hb level of 6.63 g / dl. Thrombocytopenia was associated in all cases and severe in 4 children (PQ <50,000) with an average PQ of 70,400 / mm³. Leukopenia was detected in 9 cases with an average GB of 2540 / mm³.

A hemostasis assessment made of TP +/- TCA performed in all our patients returned disrupted with prolonged TP in 8 cases (80%), with a median value of 56% and extremes ranging from 44% to 83%. A liver assessment performed in all our patients returned normal in 80% of cases, it was pathological in a single case (Case 4), combining cytolysis with mild cholestasis.

Proteemia in all patients was normal in 100% of the children. It was supplemented by an EPP indicating a hypo-albuminemia and a beta-gamma globulin block in one observation.

Hepatitis B and C serology returned negative in all patients. A medullogram performed in 5 children returned to a normal marrow with no evidence of myeloproliferative signs.

No thrombophilia assessment (coagulation factors ...) has been performed.

- Oesogastroduodenal Fibroscopy was performed in all patients and showed the presence of: Oesophageal varices in 100% of cases;
- Grade I: in 2 cases associated with VO grade II (obs.8) and grade III (obs.3)
- Grade II: in 5 cases associated with VO grade III in 2 cases (obs. 4 and 10)
- Grade III: in 3 cases
  - Grade I gastric varices in 2 cases.
  - Hypertensive gastropathy in 5 cases.

Doppler ultrasound was performed in all patients, and was completed in 2 cases by preoperative angio-scanning Case 3 and 4) to better explore portal vasculature.

In 100% of the cases, the Doppler ultrasound revealed the diagnosis of an HTP on portal cavernoma through: Direct signs of portal-portal vasculature which are: multiple anechoic formations and peri-liver CVC in the hepatic hilum, with impermeable or stenotic portal trunk in 2 cases and diffuse portal thrombosis in 1 case. And indirect signs that are: a vesicular wall thickened in 8 cases and a small epiploon thickened in 5 cases.

Hepatobiliary exploration found a heterogeneous echogenicity liver in all patients, hypertrophic in 3 cases, atrophic in 2 cases and dysmorphic irregular contours in 5 cases, with a segment I or caudate lobe hypertrophied in 5 cases associated with a patient with a hypotrophy of the right lobe (Case 9). The intra- and extra-hepatic bile ducts were not dilated in any patient.

- In 100% of cases, homogeneous splenomegaly was noted.
- The splenic vein was dilated in 7 patients.
- The condition of the superior mesenteric vein was noted in 3 cases, including 2 cases in the duplex ultrasound and one case in the angio-scanner, it was dilated with an average diameter of 10.9 mm.
- Collateral venous circulation was found in 8 cases. The histological examination focused on the liver with 5 histological samples taken using a preoperative liver biopsy puncture in one case (Case 7) and a biopsy of the liver peroperatively in 4 cases, thus demonstrating:
  - Signs of portitis with minimal congestive fibrosis in 3 cases.
  - Signs of portitis with cirrhosis in one case: grade F4 fibrosis and grade A1 activity (observed on liver biopsy)
  - Signs in favor of congenital biliary fibrosis without cirrhosis, in one case.

The histological study of the spleen was performed on the 3 pieces of splenectomy (Cases 5, 6 and 9) and found a fibro-congestive spleen.

8 cases in our series received surgical treatment, compared with 2 who received esophageal varicose vein sclerotherapy performed exclusively in
Patients and splenectomy alone in 3 cases of almost one case per 2 years, which represented 8 children underwent surgery including portocaval anastomoses in 5 patients and splenectomy alone in 3 other children. 2 children in our series received endoscopic treatment consisting of OV sclerotherapy, (Cases 8 and 10). Portocaval anastomoses consisted of:
- 1 Warren intervention: terminolateral distal spleno-renal anastomosis with preservation of the spleen and ago-portal disconnect (Case 1).
- 1 end-to-side distal spleno-renal anastomosis with conservation of the spleen without ago-portal disconnection (Case 7).
- 1 endocervical spleno caval anastomosis (Case 3).
- 2 interventions of Drapanas: mesenteric-vena cava anastomosis with interposition of jugular graft (Cases 2 and 4).

**And 3 splenectomies (Cases 5, 6 and 9)**

The immediate operative follow-up is simple except for one patient (Case 2) who has undergone the appearance of ascites which has been dried up after one month of diuretic treatment and sodium-reduced diet.

For the distant suites it was possible to obtain a recoil of variable duration except for a patient (Case 9) which was unfortunately lost sight of.

The clinical course was good in general with absence of haemorrhagic recurrence in 8 patients, regression of SMG in 2 cases, and disappearance of abdominal pain in the 3 patients who presented it, and the appearance of a sensitivity of l right hypochondry in a patient (Case 10).

Only one patient, who underwent splenectomy and was placed on aspirin, had a low abundance haematemia at one month of the intervention (Case 5). An OGDF was performed, accompanied by sclerotherapy of four oesophageal varices. This conduct allowed the eradication of bleeding recurrence with a decline of 6 months.

**An OGDF realized in the case n ° 3 objectified the virtual disappearance of the OV**

Control hemograms were performed in 6 cases and showed the disappearance of hypersplenism in 5 cases and its persistence in a patient (Case 10) in which the conduct held consisted of a simple ligature of the OV.

Thrombocytosis was noted in 2 of 3 splenectomized patients (Cases 5 and 6), who regressed after 6 and 3 months, respectively.

**An abdominal ultrasound scan (+/- Doppler) was performed in 8 of the 10 patients in our series and found**

In the 5 patients who had undergone surgery with anastomosis: 3 patients had a permeable anastomosis, in 1 of the patients, the Doppler echo showed a thrombosis carried outside the anastomosis, appeared 1 month after the intervention, having disappeared after anticoagulant treatment.

**Ultrasonography was not performed in 1 patient (Case 2)**

In the 3 patients who had undergone splenectomy: Doppler ultrasound revealed permeable portal branches without visualization of the portal flow in 1 patient. Thrombosis of the portal vein, splenomorsaraic trunk and the right portal branch appeared.

1 month after the intervention, having disappeared on a control ultrasound after anticoagulation treatment Ultrasound was not performed in 1 patient (Case 9). In the 2 patients who had undergone OV ligation, ultrasongraphy revealed respectively perioral and peri-splenic CVC (Case 8) and peri-gastric and splenorenal vesicular CVC (Case 10).

**DISCUSSION**

Portal cavernoma is the result of chronic occlusion of the extrahepatic portal system. It is formed by a network of veins whose caliber, initially millimetric or microscopic, is increased and within which travels a hepatopete portal blood. In children, portal cavernoma is a major cause of portal hypertension called "pre- or infra-hepatic" or "extra-hepatic" [1]. Its incidence varies from one series to another. In our study, we collected 10 cases in 18 years, an incidence of almost one case per 2 years, which remains a low figure in comparison with other series [1]. The portal cavernoma occurs at any age with an average of 10 to 30 years depending on the different series [2,3] The age in our series does not differ from those recorded in the literature, with a median of 10.7 years to the discovery of the disease. The portal cavernoma does not seem to be a sex-related pathology. On the one hand, studies [2, 4,5], noted a male predominance with, respectively, a sex ratio of 2.3 / 1.2 / and 2. On the other hand, the other series [1,3] noted a predominance of the female sex with, respectively, a sex ratio of 2 and 1.2. In our series, there is no predominance with a sex ratio of 1. No malformation was observed in our patients, which is consistent with their rarity in the literature. An umbilical catheterization ATCD for neonatal resuscitation exposes to portal thrombosis. None of our patients had undergone this gesture during her neonatal period. Umbilical...
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Umbilical catheterization for the resuscitation of a premature infant has been recorded in a series [1]. An ATCD of abdominal trauma, whether accidental or surgical, could be responsible for abdominal thrombosis. Similarly, a repeated venous thrombosis or at a young age in the family could lead to the diagnosis of a thrombotic state or familial thrombophilia. None of these ATCDs were noted in our series. An ATCD of abdominal infection regardless of its location could be responsible for abdominal thrombosis. We did not notice any of them in our series. Haemorrhages, most often digestive, are the most frequent reason for consultation and the percentage varies according to the series 80%, 77% and 90% of cases [1-3]. It is mostly haematemesis followed by epistaxis.

Splenomegaly is the second most common reason for consultation in children. It reveals the disease in 22% [2] and 9% [1] of the cases according to the series. In our study, hemorrhages are revealing in 70% and SMG in 30% of cases. The clinical signs are represented by the haemorrhages digestive type of hematemesis. These upper digestive haemorrhages are related either to the rupture of varices (oesophageal, cardio-tuberository) or bleeding hypertensive gastropathy, either spontaneously or following a drug (NSAID or aspirin). They are favored by coagulation disorders often present because of thrombosis and hypersplenism [6].

These haemorrhages, which are represented by haematemesis, are often followed by melena or rectorrhagia by rupture of ectopic varices, particularly in the colon and rectum (haemorrhoids). We note that hemorrhoids were not searched in any case in our series and rarely reported in the literature (2%) [6]. There is also a frequency of extradigestive hemorrhages made of epistaxis, gingivorragies, purpuric lesions ... explained by thrombocytopenia and existing disorders of coagulation. In our series, we recorded epistaxis in 2 cases. The frequency of haemorrhages in our series is 90%, made of hematemesis in 70% of cases, which joins the data of the literature: 80% [2], 77% [3], 46% [7], 40% [8] and 90% [1].

SMG is a common sign that patients present with either extra or intrahepatic HTP. It should always lead the clinician to think of a possible HTP in question, especially when it is associated with haematemesis. It may be asymptomatic for incidental discovery during a general clinical examination. As, it can be found in the exploration of abdominal pain including the left hypochondrium often related to a splenic infarction. It can take considerable dimensions causing distension and discomfort, sometimes exceeding the umbilicus and filling the iliac fossa. Usually responsible for hypersplenism, it would only aggravate hematemesis and epistaxis episodes. In our series a SMG was present in 100% of the cases. Similar results have been found in the literature; more than 80% for Leger [2], 100% for El Bouazzaoui [3] and 93% for Dibi [1].

The collateral venous circulation is of upper abdominal topography and lower thoracic and in a direction of updraft. This is a main sign of a PH, however it is not constant. It was present in 15% for Leger [2], in 55% for El Bouazzaoui [3] and in 45% in the Dibi series [1]. In our series, a CVC was found in 50% of cases. Ascites is rare in children with portal cavernoma and remains moderate and transient. It is present in 24%; 33% and 28% of cases according to different series [1-3]. In our series, ascites was not found in any case. The abdominal pains are of variable seat, epigastric, of the left hypochondrium. These abdominal pains can be intense and atypical in a non-specific gastrointestinal tract sometimes in a febrile context related to acute or prolonged port thrombosis. Hepatomegaly was found in our series in 30% and for other series HMG was noted respectively in 32%, 22%, 7% of cases [1-3]. Jaundice can occur in these children with portal cavernoma, often following a bleeding episode but exceptionally related to portal biliopathy. As the literature studies show, it is rare in children [9,10].

Jaundice was absent in our series and in the other series [2] was observed in 10%; and in 11% of cases [3, 1]. Fever was present in 11% of cases and 37% for series reported in the literature [1, 2]. We noted in our series 2 febrile cases. The failure to thrive caused by extrahepatic HTP remains controversial. Biology plays a key role in the diagnosis of extra-hepatic HTP by asserting the integrity of liver function and clearly reflects the consequences of this portal obstruction, showing hematological and hepatic repercussions. It also makes it possible to detect hereditary or acquired abnormalities of pro-thrombotic or anticoagulant factors in favor of thrombophilia. Esophagogastroduodenal fibroscopy (FOGD) has become an inevitable examination for any gastrointestinal bleeding. It is indicated for diagnostic, prognostic and therapeutic purposes.
In the context of a HTP, an OGD Fibroscopy makes it possible to specify the presence or absence of oesophageal varices, gastric and hypertensive gastropathies, as well as their grade and the haemorrhagic risk, according to the modified classification of Stringer and Schwartz [11-13]. In the literature, oesophageal varices (OV) were visualized in the lower third of the oesophagus, for the different series in 97%; 96%; 95% and 62% of cases [1, 3, 7, 14]. Gastric varicose veins are less frequent, with mainly cardio-tubercular siege and are considered to be at high risk of haemorrhage. They were found in the series with percentages of 25%; 23% and 12% of cases [1, 3, 7]. In our series, VOs were found in 100% of cases; gastric varices in 20% of cases; Hypertensive gastropathy in 50% of cases and no duodenal varices. Doppler-coupled ultrasound is the first-line examination for the discovery of digestive clinical signs; and remains an excellent means of diagnosis or even for a routine abdominal exploration and monitoring of a portal cavernoma. In our series it made it possible to pose the diagnosis of portal cavernoma in 100% of the cases as in the other series [1, 15]. Scanning also makes it possible to diagnose portal thrombosis. The CT offers the possibility of views in coronal or sagittal planes which facilitates the visualization of the thrombus on the entire portal system. This examination was performed in 2 of our patients and confirmed the diagnosis of portal cavernoma in both cases with precise description of the mesenteric and splenic vein diameters.

The goal of treatment is to eliminate the risk of bleeding without adding risks of complications specific to different therapeutic modalities. The essential treatment for hematemesis is transfusions. They will be administered according to the abundance of bleeding and the hemoglobin and hematocrit levels and the stoppage of bleeding by direct tamponade or endoscopic treatment by ligation or sclerotherapy. In children, direct measurements of the effect of β-loquant demonstrated a reduction in splenic pressure when the cardiac index was reduced by 25%, however, there were no randomized studies using β in children with HTP for primary prevention of OV [16]. The surgical treatment of HTP in children has really evolved over the last decade. This surgery can be done in three ways: Non-derivative interventions; The porto-cave derivations and which aim to lower the portal pressure by circumventing the obstacle, pre-hepatic in our context and the mesenteric-Rex bypass or portal reperfusion which is a more physiological technique, and very promising in the healing treatment of portal cavernomas . [17-20]. In our series endoscopic treatment was used in 20% of cases; however, in 80% of cases, the children received surgical treatment.

Three interventions are currently recommended in the treatment of portal cavernoma: Warren-type distal splenorenal anastomosis, which is a selective diversion without splenectomy and requires a permeable, good-sized splenic vein, mesenteric-cave anastomosis by graft interposition. type Drapanas that can be made in small children, and the meso-Rex anastomosis or portal reperfusion connecting the VMS or another vein of the trunk carries, to the left portal branch at the recess of Rex using a graft; technique that helps restore normal physiology.

Drapanas’ intervention appears to be the most appropriate for emergency situations when portal diversion is feasible [21,22]. In the opposite case, devascularization by oesophageal transection or ligating of the VO with clips stops the bleeding [23,24].

When hemorrhage occurs after a bypass, first recanalize a thrombosed shunt or dilatation with percutaneous angioplasty. Indeed, a study in Bicêtre showed the effectiveness of this procedure [11]. On the other hand, if this method proves to be inefficient, it is recommended to carry out another more central derivation or to realize a partial or total aza-portal disconnection.

CONCLUSION

Portal cavernoma is often responsible for portal hypertension in children secondary to extra-hepatic block by often thrombotic occlusion of the portal vein. This is the expression of the association of a local cause, traumatic or infectious, with another, general: prothrombotic pathology that must be sought.

Clinically, portal cavernoma is manifested by extrahepatic HTP signs made of recurrent haemorrhagic episodes by VO rupture, splenomegaly and hypersplenism. In addition, the diagnosis of cavernoma is based on imaging and especially Doppler ultrasound or angio-scanner, sensitive and insignificant exams that have replaced invasive exams.

Biology, on the other hand, most often discovers hypersplenism. Oesogastroduodenal fibroscopy is the basic examination to perform in case of hematemesis or even signs of portal hypertension. This is an examination that dictates the therapeutic behavior.

The new surgical procedures have significantly improved the management. In fact, the interventions that allow the decompression of the portal network have made it possible to correct the complications of the HTP and to improve the quality of life, whereas the nonsurgical treatment is symptomatic and often requires a long monitoring and a regular recurrence of the endoscopic sessions, to be able to eradicate varicose veins that recidivate. However, hemorrhage during the procedure is a risk, which is why non-surgical treatment is recommended before any surgery.

Finally, it is concluded that the portal cavernoma is a crippling pathology of the child who
suffers from it requiring a high level of multidisciplinary care between pediatricians, pediatric surgeons, hepatogastroenterologists and radiologists, in order to make an early and appropriate decision.

REFERENCES
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