Portal vein aneurysm (PVA) is a rare vascular malformation (0.067% incidence) [1], representing fewer than 3% of all venous aneurysms [2]. First described by Barzilai and Kleckner in 1956 [3,4], almost 150 cases are now reported in literature [5] (only 68 cases in English) [6]).

Morphologically, portal vein aneurysm can be fusiform or saccular [7]. Two main categories have been described: intrahepatic or extrahepatic. Extrahepatic aneurysms are more common, located at the main portal vein in 52% of cases, at the superior mesenteric–splenic vein confluence in 44%, and rarely (3% to 4%) at the level of the right or left portal branches [4]. Portal vein diameter being variable, an aneurysmal dilatation of the PV is defined according to the venous upper limit of range for age [8–10]. Intrahepatic aneurysms and those involving the umbilical portion of portal vein are rare with only few cases being reported [11–13].

Clinical presentation varies, especially in relation to the aneurysm size. The majority of patients presenting with nonspecific abdominal pain (44% to 54%), while many others are asymptomatic and diagnosis is incidental (25% to 38%). Rarely, patients are diagnosed with portal hypertension or gastrointestinal bleeding (7.3% to 9.8%) [3,4]. In this report, 3 children with pre-hepatic portal hypertension and intrahepatic portal aneurysms are presented, and a surgical cure is described.

### 1. Patients and methods

Between January 2007 and December 2011, three children with PVA were identified out of a population of 105 children who were radiologically evaluated for extrahepatic portal hypertension (3%, all 3 female). Patient characteristics, clinical presentation, anatomy of the PVA, surgical correction and outcome were reviewed. Anatomical assessment of the anatomy was done by standard imaging (Doppler Ultrasound (US), Angio CT, Angio MRI) and transjugular retrograde portography (gradient measurement and retrograde portography) in all cases.

#### 1.1. Patients

#### 1.1.1. Patient 1

At the age of 18, she was admitted to our centre after a single episode of massive variceal bleeding a month earlier. Endoscopy had shown four F2 varices with red stripes in the lower portion of the

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**Prehepatic portal hypertension with aneurysm of the portal vein: Unusual but treatable malformative pattern**

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**ABSTRACT**

Introduction: Portal vein aneurysms (PVAs) are usually located at the vein trunk or at its bifurcation, rarely intra-hepatic, or at the umbilical portion. Etiology remains unclear.

Methods: Three children with PVA were identified over a 5-year period. PVA anatomy was assessed by Doppler Ultrasound, Angio CT/MRI, and trans-jugular retrograde portography.

Results: Three children with intrahepatic PVA (including the umbilical portion) were identified during assessment for pre-hepatic portal hypertension: all had splenomegaly and hypersplenism. One presented with massive variceal bleeding. In two cases, a portal vein cavernoma was found, and in the third a severe stricture at the portal bifurcation was observed. Restoration of portal venous flow was achieved by a meso-Rex bypass in two cases and transposing the PV into the Rex in one. High hepatopetal portal flow was restored immediately, with follow-up confirming long-term patency and resolution of signs of portal hypertension with time.

Conclusions: These original observations suggest a common initial malformative pattern consisting of a portal venous stricture/web causing a post-stenotic aneurysmal dilatation of the intrahepatic portal branches complicated by thrombosis and cavernomatous transformation of the portal vein trunk. Importantly, the Meso-Rex bypass allows restoring a normal portal flow and cures the portal hypertension.

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esophagus, and she was treated by elastic bands positioning. She had splenomegaly (US span: 20 cm), thrombocytopenia (platelet count: 57,000/ml) and normal hepatic function tests. At radiological assessment, a portal cavernoma and aneurysm of both right and left portal branches, including a huge aneurysmal transformation of the Rex recessus, were observed.

1.1.2. Patient 2

A 5-year-old girl presented initially with variceal bleeding; esophageal varices, factor VII deficiency, splenomegaly (US span: 14.5 cm) and thrombocytopenia (platelet count: 86,000/ml). At imaging, a malformative pattern combining PVA of both portal branches extending into the Rex recessus, a normal extrahepatic portal vein trunk and a severe stricture of the main portal trunk at the level of the bifurcation were found (Figs. 1 and 2A). An attempt at percutaneous transhepatic angioplasty/stenting of the stricture in another centre failed, and she was referred to our centre for surgical treatment.

1.1.3. Patient 3

Splenomegaly and portal vein cavernoma were incidentally diagnosed in a 3-year-old girl with an uneventful history and absence of previous gastrointestinal bleeding. Endoscopy showed esophageal varices F2, gastric varices and mild hypertensive gastropathy. She was in good general conditions, with splenomegaly (US span: 16 cm), thrombocytopenia (platelet count: 56,000/ml) and normal hepatic function tests. Radiological assessment showed a portal cavernoma with patent intrahepatic portal branches of a normal diameter, and an aneurysmal transformation of the Rex recessus (Fig. 2B).

1.2. Details of radiological assessment

The Rex recessus was aneurysmal in all three cases, with the aneurysm involving the other main portal branches in two. In none was the extrahepatic portal main venous trunk found dilated. In 2 cases the latter trunk was thrombosed with secondary cavernomatous transformation. Patient 2 was remarkable with a patent portal vein and a severe stricture immediately below the bifurcation (Fig. 2A). In all cases, splenic vein, mesenteric vein, and their confluence, were normal.

The maximum diameter of Rex recessus, as measured at Doppler US, ranged from 14.58 mm to 20.20 mm (mean 17.62 mm). All PVAs had a fusiform shape, and at US typically presented as large anechoic structures (Fig. 1B). Colour Doppler US and duplex Doppler US showed the characteristic aspect of venous structures with constantly rotating blood flow within the lesion and its typical spectral analysis pattern in patients with high flow [8,12,14,15]. Flow in liver-aneurysms was turbulent and velocities at that level ranged from 25 cm/s to 70 cm/s (mean 50.3 cm/s). On CT, the aneurysmal segments appeared as a well-circumscribed enhanced structure as part of the intrahepatic portal venous system (Fig. 2B). On T1 and T2 weighted MRI images, PVA appeared hypointense. More detailed anatomical assessment of the intrahepatic portal system was obtained in these patients by 3-dimensional CT reconstructions (Fig. 2A), and direct or indirect angiographies: this clearly delineated the aneurysmal transformation and its relation with the Rex recessus and the normal segmental portal vein branches.

1.3. Management

All three patients were successfully managed surgically by the same senior surgeon: a meso-Rex bypass was performed in two, using the left internal jugular vein as a bypass (the technique has been previously described [16]). In the third case, the portal vein was freed and divided below the bifurcation: at that point, it appeared technically easier to mobilize it anteriorly and anastomose it directly, end-to-side, into the dilated Rex recessus.

In all 3 patients, a high blood flow – from 40 cm/s to 50 cm/s (mean 48 cm/s) – was recorded intra-operatively within the bypass (or the portal vein in case 2), with excellent flow restored within the whole liver (Fig. 3). Current follow-up ranges from 14 to 24 months (median ± SD: 18 ± 5 months; mean 18.67 months). Clinical and radiological assessments have confirmed the good outcome in all 3 patients with resolution of portal hypertension symptoms. At follow up Doppler US, the bypass flow ranged from 30 to 50 cm/s (mean 43.33 cm/s) and the aspect of aneurysm significantly normalized with a mean diameter of 15.30 mm (ranging from 13.52 mm to 18.85 mm) and a mean flow of 29.33 cm/s (ranging from 25 cm/s to 33 cm/s).

The platelet count increased with time (Table 1) and the spleen is clinically no longer appreciable (spleen span at US decreased to a mean of 13.5 cm). Coagulation tests tended to normalize with time in all three patients (Table 1). No endoscopic control has been done after the operation, as we do usually after successful Meso-Rex bypass.

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**Fig. 1.** (A) Aneurysm of the Recessus of Rex (Patient II): aspect at conventional angiography (venous phase of superior mesenteric artery angiography). (B) Ultrasound aspect of an aneurysm of the right and left portal branches, typically presenting as a large anechoic fusiform structure (Patient II).
when the clinical condition improves and if excellent blood flow is measured in the bypass.

2. Discussion

The portal vein is the most common site of visceral venous aneurysms and can present as a focal saccular or fusiform dilatation [8,17,18]. Precise etiology is still a matter of debate with investigators favoring two hypotheses.

Embryogenesis and formation of the normal portal system are the results of a complex process initiated by formation venous systems (umbilical and omphalomesenteric – or vitelline – veins) bilaterally (right and left), followed by their interconnections (R–L anastomoses) and ending in the creation of the usual portal system by a final process of involution/regression of parts of the former venous system (http://www.embryology.ch/anglais/pcardio/venen02.html and http://fr.slideshare.net/anathatiger/embryology-vascular-development). The “congenital” theory postulates that incomplete regression of some vein or a variant branching pattern of the portal vein may determine aneurysmal transformation of the portal vein [3,19] or of the umbilical portion of the left portal vein [11].

For the supporters of the “acquired” theory, PVAs are the consequence of a weakening of the venous walls due to acute pancreatitis, or trauma, or even a consequence of a portal hypertension secondary to liver disease [3,4,19]. Not to mention that the “weakened venous wall hypothesis” has also been considered by the supporters of the congenital theory as a possible congenital alteration of the venous wall which could cause subsequent venous dilatation under normal portal pressure [15]. Although portal hypertension is generally accepted as a cause of acquired portal vein aneurysm [12,14], the latter is rather a moderate dilatation of a patent portal vein trunk, then a true aneurysm being the consequence of a high splanchnic flow associated with hyperdynamic circulation in the cirrhotic patient. A similar effect on portal vein diameter is observed in patients with normal liver and a high portal flow secondary to arterio-portal fistula or porto-hepatic shunting [20,21]. Remarkably, in our three cases, no loco-regional abnormal condition was found and the liver was normal: there is no reason to believe that portal flow was increased as a primary cause.

Overall, the various theories proposed for the genesis of PVA offer a reasonable explanation for cases with dilatation of the main portal vein, with or without venous abnormal anatomy, but not a clear explanation for those aneurysms that are located higher and especially the intrahepatic types. Also, it does not clearly explain the association between aneurysm and upstream portal cavernoma in young children.

In two of our patients, the portal vein trunk was thrombosed with the aneurysm located downstream at the level of the right and left portal branches. In fact, the relation between PVA and venous thrombosis, not uncommonly seen in cirrhotic patients (up to 30 % [14]), is a “chicken–egg” debate. Turbulence or areas of blood stasis within an aneurysm could trigger thrombus formation. In these
patients, thrombosis is a consequence, rather than the cause of the malformation, and is then located at the site of the aneurysm, not upstream as in our cases.

Interestingly, Kimura et al. have described in patients with thrombophilia, the formation of thrombus that may grow into an incomplete membrane: this process could lead not only to thrombosis but also to the formation of a membrane within the vein [22,23].

Patients 2 and 3 had very similar transformation of the portal venous system, the latter patient having a cavernomatous transformation of the portal vein, and the former a patent portal vein trunk with a severe stricture at the vein bifurcation. The latter stricture is evidently the cause of a typical post-stenotic dilatation of the vein. The PVA anatomy of both cases is so similar that it suggests that patient 3 had initially a similar portal vein stricture to that of Patient 2, complicated later by secondary thrombosis of the portal trunk and cavernoma formation at its level.

According to Kimura et al., a thrombotic origin of the stricture cannot be ruled out.

Interestingly, the association of intrahepatic PVA and extrahepatic portal hypertension caused by thrombosis of the portal vein trunk, in the presence of a normal liver and in the absence of extensive splanchnic venous thrombosis, makes that particular condition amenable to a cure by surgically restoring the portal system continuity. In these patients, it was achieved by direct transposition and anastomosis of the portal trunk into the dilated Rex recessus in one case, and by creating a bypass between the mesenteric vein and the Rex recessus (bypass Meso-Rex) in the two patients with portal cavernoma. The technical aspect of that procedure, as well as the fact that it respects the physiology of portal venous return through the liver, has been emphasized previously [16]. In the 3 patients, the procedure has been successful and associated with the relief of symptoms and complications caused by portal hypertension.

Table 1
Changes in spleen span and hypersplenism: before (pre) and after (post) meso-Rex bypass.

<table>
<thead>
<tr>
<th>Patient</th>
<th>FU months</th>
<th>Pre</th>
<th>Post</th>
<th>Pre</th>
<th>Post</th>
<th>Pre</th>
<th>Post</th>
<th>Pre</th>
<th>Post</th>
<th>Pre</th>
<th>Post</th>
<th>Pre</th>
<th>Post</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>18</td>
<td>20 (3)</td>
<td>14.5 (0)</td>
<td>57</td>
<td>105</td>
<td>31.5</td>
<td>27.6</td>
<td>16</td>
<td>13.9</td>
<td>1.26</td>
<td>1.01</td>
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<td></td>
</tr>
<tr>
<td>Patient 2</td>
<td>24</td>
<td>14.5 (4)</td>
<td>12 (0)</td>
<td>86</td>
<td>135</td>
<td>35.8</td>
<td>31.2</td>
<td>17.8</td>
<td>15.8</td>
<td>1.51</td>
<td>1.32</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patient 3</td>
<td>14</td>
<td>16 (5)</td>
<td>14 (0)</td>
<td>56</td>
<td>142</td>
<td>36.5</td>
<td>31.9</td>
<td>15.8</td>
<td>12.6</td>
<td>1.25</td>
<td>1.00</td>
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</tr>
</tbody>
</table>

Abbreviations: FU: Follow-up; PLT: platelet count (million/ml); PTT: Partial thromboplastin time (seconds); PT: Prothrombin time (seconds); INR: International Normalized Ratio.

* Spleen span was measured in cm, as maximum span at ultrasound imaging (below rib margin at clinical examination.)
3. Conclusions

These three observations strongly support the hypothesis that a stricture, either caused by congenital stricture or partial thrombosis of the main portal vein is the primary lesion, with aneurysmal transformation being secondary to the stricture in the portal vein (as observed in case 3 and suspected in case 2). This produces turbulences and aneurysmal transformation.

As a third event, complete thrombosis of the portal vein trunk may occur with cavernomatous transformation being the last event; this sequence would then explain easily that, in some patients, PVA can be found with complete thrombosis, the latter occurring at a later stage as a last complication and causing in turn a cavernomatous transformation of the portal main trunk.

It is interesting to note that aneurysmal transformation of the portal vein, per se and in the absence of secondary complications (i.e., thrombosis) is not clinically relevant and, per se, does not need treatment. In contrast, prehepatic portal hypertension, presented here, as a secondary is the cause of delayed morbidity and can be cured by appropriate intervention. This series shows that the MesoRex bypass can offer a cure for those patients when the liver is normal.

References