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Liver Transplantation from Donors with Down Syndrome

Benjamin Fernandez^{1,2}, MD; Chetana Lim¹, MD, PhD; Claire Goumard^{1,3,4}, MD; Fabiano
Perdigao¹, MD; Geraldine Rousseau¹, MD; Eric Savier¹, MD; Alessandra Mazzola⁵, MD;
Filomena Conti^{3,4,5}, MD, PhD; Olivier Scatton^{1,3,4}, MD, PhD.

1. Department of Hepato-Biliary and Pancreatic Surgery and Liver Transplantation, AP-HP Pitié-Salpêtrière Hospital, Paris, France
2. Department of Hepato-Biliary and Pancreatic Surgery and Liver Transplantation, Haut Lévêque Hospital, CHU Bordeaux, France
3. Sorbonne Université, UMPC Université Paris 6, Paris, France
4. Centre de Recherche de Saint-Antoine (CRSA), INSERM, UMRS-938, Paris, France
5. Department of Hepatology, AP-HP Pitié-Salpêtrière Hospital, Paris, France

Corresponding author and reprint requests:

Prof. Olivier SCATTON

Department of Digestive, Hepato-Biliary and Pancreatic Surgery and Liver Transplantation
AP-HP Pitié-Salpêtrière Hospital, Paris, France

Email: olivier.scatton@aphp.fr

Telephone: +33184827427

To the Editors:

The current organ shortage has resulted in extended criteria for donor selection. The use of risky cadaveric liver donors, most of which are older, obese or donation after circulatory death donors has been developed to face the growing organ demand. As improvements in medical care for patients with Down Syndrome (DS) have led to an increase in their life expectancy, the question of potential liver donors with DS should be considered. Because congenital vascular malformations of the liver have been described in patients with DS, LT using a graft from a donor with DS may be challenging. We report here, for the first time, two cases of successful cadaveric liver transplantation (LT) from donors with DS.

Case series

The first recipient was a 38-year-old man with alcoholic and metabolic syndrome cirrhosis who underwent orthotopic LT. The donor was a 28-year-old man with a history of DS who died from a post-traumatic intracerebral bleeding. Previous medical history included thyroidectomy and cholecystectomy. Laboratory explorations showed: total bilirubin = 7 $\mu\text{mol/l}$, aspartate-aminotransferase = 24 IU/l, alanine aminotransferase = 20 IU/l, γ -glutamyltransférase = 89 IU/l, alkaline phosphatase = 136 IU/l, prothrombin time = 72%. Computed tomography revealed a complete stenosis of the left branch of the portal vein and an atrophy of the left lobe (**Figure 1A**). The right branch of the portal and the portal vein remained patent. No other cardiac and vascular abnormalities were found. Caval reconstruction was performed using the Piggyback technique. Portal reconstruction was performed using portoportal anastomosis. Arterial and biliary reconstructions were performed as usual. The cold ischemia time was 480 minutes. Anticoagulation including low-molecular-weight heparin as a prophylaxis against the extension of portal vein thrombosis and aspirin as a

prophylaxis against arterial thrombosis was started in intensive care unit. The patient underwent relaparotomy for intra-abdominal bleeding on postoperative day 12, but no bleeding spot was identified. Two months after LT, the patient is alive with normal liver function and persisting left portal vein stenosis (**Figure 1B**).

The second recipient was a 33-year-old woman with a history of Kasai procedure for biliary atresia. She developed secondary biliary cirrhosis. The donor was a 47-year-old man with a history of DS who died from an acute obstruction of his permanent tracheostomy. Previous medical history included permanent tracheostomy for severe laryngeal papillomatosis.

Laboratory explorations showed: total bilirubin = 9 $\mu\text{mol/l}$, aspartate-aminotransferase = 31 IU/l, alanine aminotransferase = 24 IU/l, γ -glutamyltransférase = 24 IU/l, alkaline phosphatase = 72 IU/l, prothrombin time = 69%. Computed tomography did not reveal any cardiac, venous, portal or arterial anomalies. The cold ischemia time was 446 minutes. Caval, portal, arterial and biliary reconstructions were performed as usual. Anticoagulation included aspirin as a prophylaxis against arterial thrombosis. Two months after LT, the patient is alive with patent vascular reconstructions and normal liver function.

Discussion

To our knowledge this is the first series of successful LTs from donors with DS, suggesting that such graft from DS should not be excluded from liver donation. While congenital cardiac anomalies and intra-abdominal malformations are common, congenital vascular malformations of the liver have been described in children with DS ¹. The latter are defined by the vascular connection between the hepatic artery, portal and hepatic veins and inferior vena cava. They may be divided into two categories according to the nature of the vascular connections (**Figure 2**): i) veno-venous connections (i.e, shunt between portal venous and systemic venous system) including portocaval shunts (shunt between the portal vein and

inferior vena cava, the so-called Abernathy malformation) and patent ductus venosus (i.e., shunt from left portal vein to the hepatic vein confluence), and ii) arterio-venous (shunt between hepatic artery and hepatic veins or inferior vena cava) and arterio-portal (shunt between hepatic artery and portal vein with subsequent risk of portal vein aneurysm) shunts. Interestingly, a novel portal vein malformation not previously described is the left portal vein stenosis, as seen in one of our two cases. In this case of congenital left portal vein stenosis, we only use additional anticoagulation as this stenosis cannot be re-vascularized with the retrograde approach through the portal vein using vascular angioplasty using balloon catheter or stenting. Moreover, performing an *in situ* or *ex vivo* left hepatectomy was judged as an excessive option with specific risks brought by the liver parenchymal surface potentially leading to bleeding or bile leak.

As for other genetic abnormalities, one case of LT using a donor with Turner syndrome (45 XO chromosomal status) has been reported in 1994 ². In this case, the donor had a replaced hepatic artery arising from the superior mesenteric artery.

Because no particular vascular malformation predominated in patients with DS, decision to use liver graft from a donor with a DS should be taken on a case-by-case basis. In the case of complex congenital connection between the arterial and portal venous system with potential subsequent arteriportal hypertension and portal vein aneurysm, liver transplant surgeons should be cautious before using this kind of graft. Indeed, these arteriportal malformations would probably need secondary embolization of arteriportal shunts, surgery with ligation of the feeding arterial vessels, segmentectomy or lobectomy ^{3,4}. In a modern era of graft assessment by computed tomography, the decision to use or not this kind of graft can be made easily. If ultrasound is used to assess the liver graft, then liver transplant teams should ask for computed tomography scan evaluation in order to seek for potential anatomical variations and shunts.

In summary, the use of a liver graft from a donor with DS for LT is safe and feasible. A meticulous assessment for vascular anomalies of the liver by computed tomography scan at least should be performed before accepting the graft.

Figure legends**Figure 1. Liver transplantation using a liver graft from a donor with Down Syndrome and a stenosis of the left portal vein.**

- A. Intraoperative view of the liver graft before procurement.
- B. Post-liver transplantation computed tomography. Note the stenosis of the left portal vein and the atrophy of the left liver in the recipient.

Figure 2. Classification of congenital vascular malformations of the liver in patients with Down Syndrome (Black circle indicates the malformation).

- A. Portal vein-inferior vena cava
- B. Portal vein-hepatic vein
- C. Hepatic artery-hepatic vein
- D. Hepatic artery-portal vein

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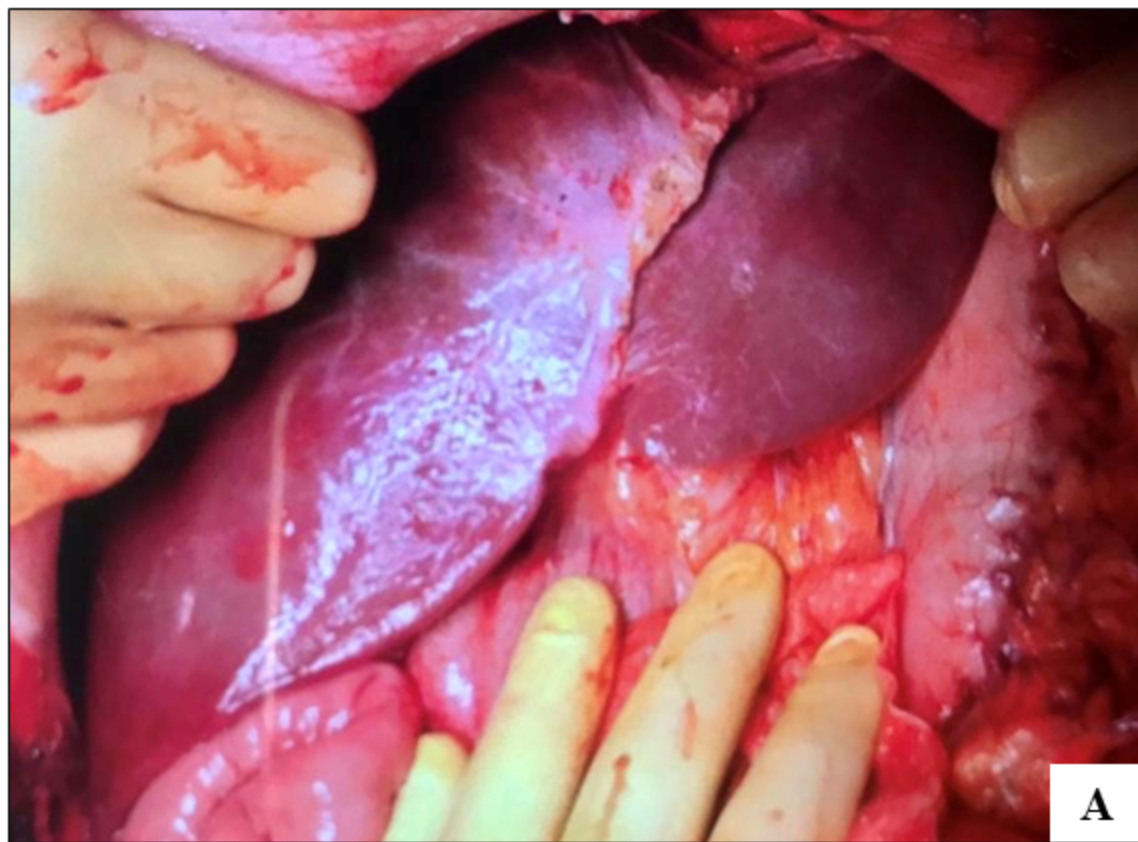
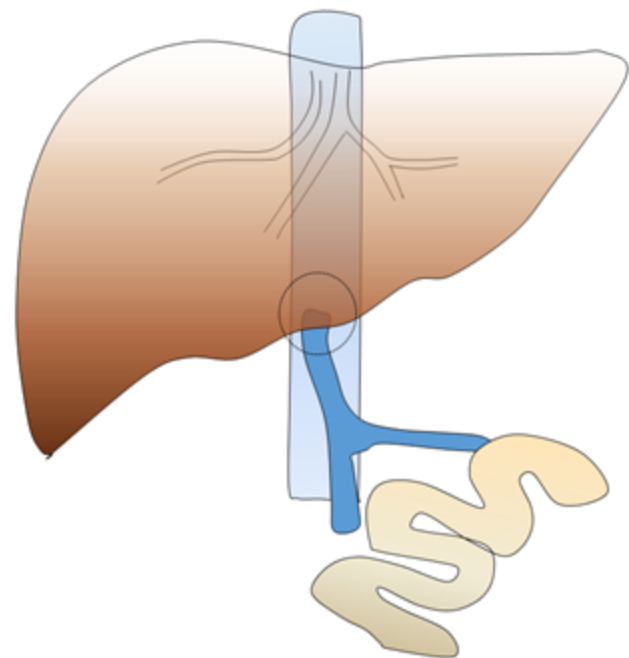
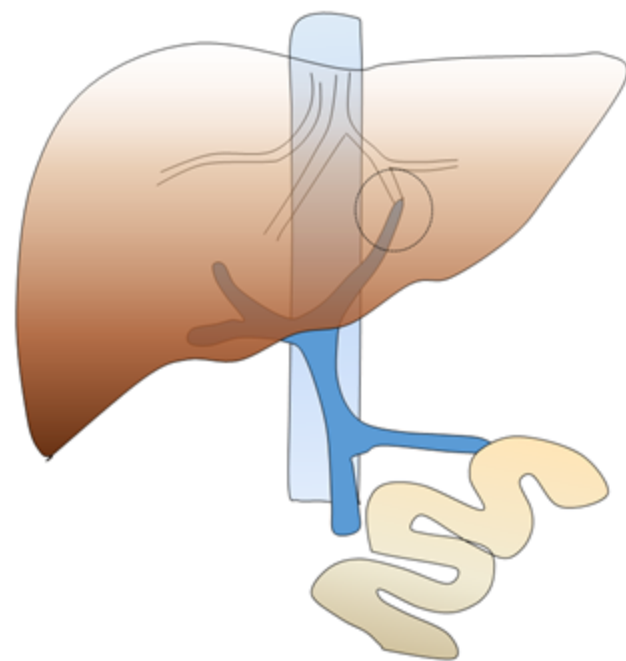
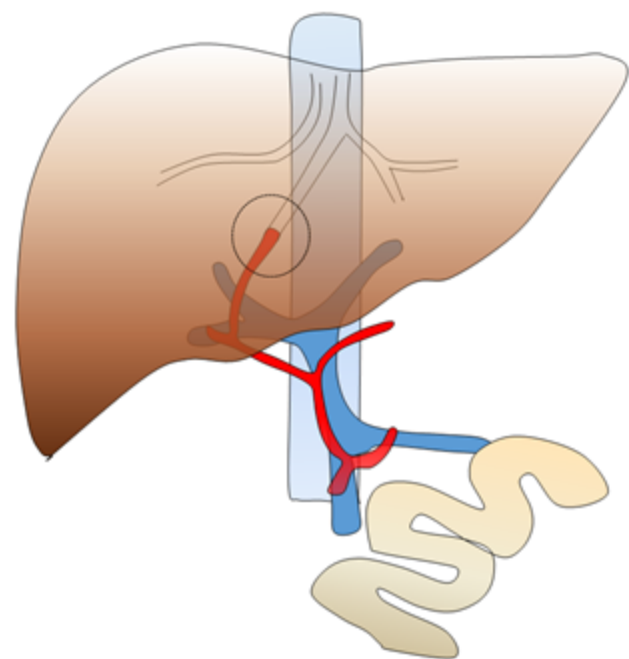
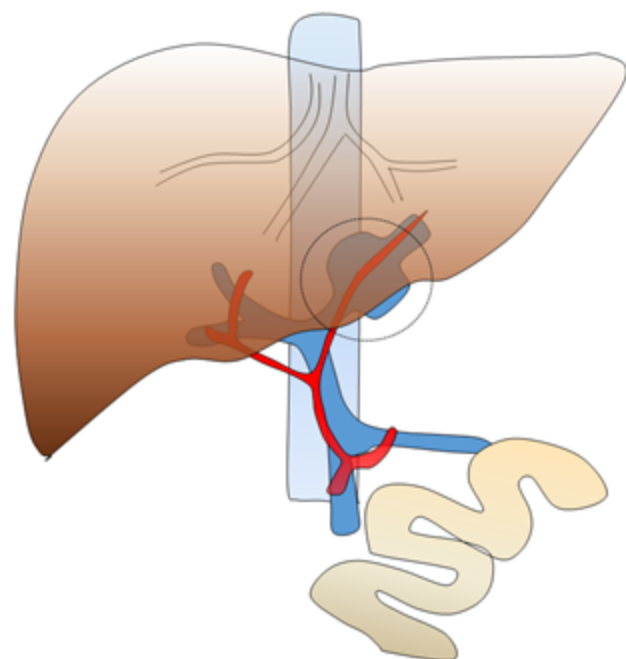


Figure 1

A**B****C****D****Figure 2**