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Modern management of portopulmonary hypertension in children: experience of an expert center

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Introduction:
Portopulmonary hypertension (PoPH) is a rare but serious complication of portal hypertension or portosystemic shunting. PoPH management may require liver transplantation (LT) but this procedure is contraindicated in patients with severe hemodynamic impairment. There are only few reports describing PoPH management and outcome in children.

Methods:
We describe our series of children with PoPH. Diagnostic workup was performed according to the current international guidelines and precapillary pulmonary hypertension was confirmed by right heart catheterization. Patient data was collected via chart review.

Results:
5 patients were identified in our pediatric center from 2013 to 2017, with a mean age of 15.8 years old (range 14-19). The underlying liver conditions were cirrhosis of unknown origin, 2 portocaval shunts, biliary atresia, and cavernoma of the portal vein. 3 patients suffered with dyspnea and 1 had no cardiopulmonary symptom. The mPAP was 42.6mmHg (range 30-54) and the indexed mean pulmonary vascular resistance (PVRi) was 8.44 Wood units (range 4.3-14). The 1st was in functionnal class (FC) III and treated by combination therapy consisting of macitentan and tadalafil. After a significant improvement of hemodynamic profiles, LT is discussed. The 2nd had no cardiopulmonary symptom. PoPH is almost resolved after shunt closure and under treatment of macitentan and tadalafil. The 3rd was in FC III and treated with macitentan and tadalafil. Shunt closure is scheduled. The 4th (biliary atresia) was in FC IV and treated by triple combination therapy with macitentan, tadalafil and IV epoprostenol. PoPH improved dramatically and the patient underwent LT. Epoprostenol was switched to oral selexipag 9 months after LT. PoPH is now controlled under macitentant, tadalafil and selexipag. The 5th underwent a surgical mesocaval shunt for a cavernoma of the portal vein. 7 years later, he presented in FC III and was bridged to liver transplantation by bosentan. LT was performed and PoPH is now resolved. **Table 1** shows patients characteristics.

Conclusions:
Outcome of pediatric PoPH in the modern era may be largelly improved when managed at an expert center offering PH and LT multidisciplinary managment. Further studies are needed to better define the pronostic and treatment in this population.

Cases (sex)	Age in year	Liver disease	Medical treatment	Surgical treatment	Hemodynamic parameters before medical treatment	Hemodynamic parameters after bridging therapy (medical treatment)	Hemodynamic parameters at last follow-up	Medical treatment at last follow-up
1 (F)	14	Cirrhosis of unknown origin	Macitentan Tadalafil	LT will be discussed	mPAP = 48 mmHg PRV = 10 WU CO = 3.5 l/min	mPAP = 35 mmHg PRV = 6.8 WU CO = 4 l/min	mPAP = 35 mmHg PRV = 6.8 WU CO = 4 l/min	Macitentan Tadalafil
2 (M)	14	Congenital portocaval shunt	Macitentan Tadalafil	Shunt closure	mPAP = 33 mmHg PRV = 4.3 WU CO = 5.6 l/min	mPAP = 22 mmHg PRV = 1.8 WU CO = 10.8 l/min	mPAP = 24 mmHg PRV = 2.6 WU CO = 6.2 l/min	Macitentan Tadalafil
3 (F)	15	Congenital portocaval shunt	Macitentan Tadalafil	Shunt closure will be scheduled	mPAP = 54 mmHg PRV = 14 WU CO = 4.9 l/min	Scheduled before closure		Macitentan Tadalafil
4 (F)	19	Biliary atresia	Bosentan Tadalafil Epoprostenol	Kasai portoenterostomy, then LT	mPAP = 48 mmHg PRV = 9.4 WU CO = 3.7 l/min	mPAP = 41 mmHg PRV = 8 WU CO = 4.3 l/min	mPAP = 33 mmHg PRV = 6 WU CO = 5.3 l/min	Macitentan Tadalafil Selexipag
5 (M)	17	Cavernous transformation of the portal vein	Bosentan	Mesocaval shunt for treatment of portal hypertension, then LT for treatment of PoPH	mPAP = 30 mmHg PRV = 4.5 WU CO = 5.5 l/min	mPAP = 22 mmHg PRV = 3.6 WU CO = 7.5 l/min	mPAP = 20 mmHg PRV = 2.4 WU CO = 5.5 l/min	None

Table 1. Patients characteristics, hemodynamic parameters and evolution