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24 year-old pregnant woman with ventricular septal defect and pulmonary hypertension

Rehak T., Nagel B., Heinzl B., Fritsch P., Köstenberger M., Gamillscheg A.
Division of Pediatric Cardiology, Department of Pediatrics, Medical University Graz, Austria

Introduction:
Pregnancy in patients with pulmonary hypertension (PAH) and congenital heart defects (CHD) is associated with a high maternal mortality. Experiences with therapeutic options during and after pregnancy are rare.

Case:
A 24 year-old woman presents to our adult congenital heart clinic in the 22nd week of gestation due to a heart murmur. She is acyanotic (SaO_2 94%), a 3/6 systolic murmur is audible and she is NYHA II. Echocardiogram shows a large VSD with bidirectional shunt. During the course of pregnancy no complications occur, Caesarean section is performed in the 37th week of gestation, she gives birth to a healthy daughter. Postpartum she receives prostacycline i.v. for 5 days, heparin as well as oxygen supplementation. 3 months later exertional dyspnoea occurs (NYHA III), cardiac catheterization shows the PA pressure systemic (PAP/SAP: 0.72) with high pulmonary vascular resistance index (PVRI) at 15 U*m^2. O_2 and NO testing results in an increased left to right shunt and a significant reduction of PVRI (3.3 U*m^2). Bosentan therapy is started, after 12 months the 6-minute walking distance (6MWD) improves from 503 m (collapse) to 560 m, PVRI decreases at rest (5.9 U*m^2) and shows sigificant vasoreagibility with O_2 testing (1 U*m^2), PAP is unchanged. Partial closure with a 6 mm fenestrated VSD patch is performed, followed by an unproblematic postoperative period. 12 months after surgery (24 months of Bosentan therapy) the exercise capacity further improves (NYHA II) and 6MWD increases to 682 m. Cardiac catheterization shows a further decrease in PVRI at rest (4.4 U*m^2) as well as a lowered PAP (PAP/SAP: 0.4). Bosentan therapy is stopped, 6 months later the 6MWD improves to 721 m.

Discussion:
Adult patients with PAH due to a large VSD without Eisenmenger physiology may show an adequate vasoreagibility of their pulmonary vasculature. Thus an uncomplicated course of pregnancy and postpartal period is possible. Bosentan may support partial VSD closure in due course and can result in an improvement of NYHA functional class and 6MWD.