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Article type: Clinical vignettes

Received: June 10, 2019.

Accepted: September 16, 2019.

Published online: September 17, 2019.

ISSN: 0022-9032

e-ISSN: 1897-4279

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Fetal and newborn percutaneous aortic balloon valvuloplasty in critical aortic stenosis followed by complex Ross-Rastan-Konno reconstruction

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Conflict of interest: none declared

Short title: Fetal valvuloplasty followed by Ross-Rastan-Konno reconstruction.

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A midgestational male fetus in the 22nd week of pregnancy was diagnosed with a critical aortic stenosis (AS) and concomitant left ventricle hypoplasia characteristic for progressive hypoplastic left heart syndrome (HLHS). To prevent heart dysfunction over the subsequent course of gestation and reverse the evolution into HLHS, intrauterine aortic balloon valvuloplasty was performed in the 26th week of pregnancy in Children’s Heart Center in Linz, Austria. The size of the balloon used for the valvuloplasty was 4mm, decompressed to 4.4mm at the pressure of 16 bar. By an uncomplicated procedure increase of fetal left ventricle ejection fraction was achieved with flow normalization both in ductus arteriosus and foramen ovale, mitral regurgitation was reversed. Right after birth, due to a recurrent constriction of aortic ostium, an urgent newborn percutaneous balloon valvuloplasty was performed. Furthermore, the procedure was redone after 15 months due to a relapse of the AS. At the age of 5 control catheterization confirmed a stable course of hemodynamically significant complex stenotic left ventricle outflow tract (LVOT) with fibroelastosis and aortic regurgitation. Concomitant losses of consciousness were a clear indication for urgent surgical treatment.

A Ross-Rastan-Konno procedure was the final step of the treatment of the 7-years-old patient after fetal and newborn-period interventions. After meticulous resection of LVOT endocardial fibroelastosis and muscular hyperplasia, top section of resected interventricular septum was supplemented with pulmonary autograft, and extensive fragment of right ventricle free wall implanted into subaortic position. The operation included vast reconstruction of the right ventricular outflow tract (RVOT) with the use of flared pulmonary xenograft. The postoperative course was uncomplicated, the boy was referred for outpatient clinic, and in mid-term follow-up is doing well.

Fetal aortic balloon valvuloplasty for congenital AS may preserve the function of left heart system and therefore prevent fatal intrauterine progression into HLHS.[1] Percutaneous interventional and surgical interventions in newborns should be a spontaneous continuation to
achieve biventricular outcome. With an optimal management of complex left ventricular outflow tract obstruction (LVOTO) the patients could continue for final feasible, modified Ross-Rastan-Konno LV preservation procedure.[1,2] HLHS presents as iterative (consists of minimum 3 surgical procedures), troublesome, long-term palliation of univentricular patients, with more than 90% mortality rate if untreated.[3] Fetal cardiac interventions for congenital heart defects such as AS with HLHS-physiology may prevent them from evolving into complete HLHS, thus improve fetal survival or even achieve biventricular outcome-like in the case of our patient. On the other hand, fetal aortic valvuloplasty still carries a 30% risk of impaired pregnancy, including fetal demise[1,4]. Precise anatomical predictors for biventricular outcome achievement are still uncertain and the results may vary among the fetuses with critical AS, likewise it depends on institutional experience.[4] Nevertheless, fetal valvuloplasty provides a chance for a left heart rescue and further therapy, including cardiac surgery such as modified Ross-Rastan-Konno procedure to improve patient’s long-time prognosis.

In summary, intrauterine cardiac intervention and successive newborn aortic valvuloplasty preserved the left heart function and initiated the maintenance that has successfully allowed for surgical LVOT reconstruction with modified Ross-Rastan-Konno procedure.

References:


Figure 1:

A. Newborn echocardiography: Complex LVOT stenosis. Fibroelastosis in LVOT (arrows). Dysplastic aortic valve after fetal balloon valvuloplasty.

B. Newborn interventional LVOT balloon valvuloplasty – level of the aortic valve (arrow).

C. Control catheterization: complex LVOT stenosis with progressive aortic valve (AV) regurgitation (arrow).

D. Postoperative echocardiography after Ross-Rastan-Konno procedure, resected
interventricular septum was supplemented with pulmonary autograft, and extensive fragment of right ventricle free wall implanted into subaortic position(arrow).