Balloon valvuloplasty for critical aortic stenosis in a fetus: a case report

Fetusta kritik aort darlığı için balon valvuloplasti: Olgu sunumu

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Summary– The mortality and morbidity of fetal aortic stenosis (AS) depend on the degree of the hemodynamic effects of the stenosis, and left ventricular (LV) adaptation, development and function during fetal life. In the case of critical AS, the development of hydrops and death *in utero* are well recognized entities. A 23-week gestation fetus was diagnosed with critical severe AS, cardiomegaly, a dilated LV with very poor contractility, and mitral regurgitation. There was a reversal of flow in the aortic arch through the ductus arteriosis and a reversed a-wave in the ductus venosus on Doppler examination. The fetus had hydrops with ascites, and massive scalp, face and skin edema. Fetal amniocentesis was normal. Aortic valvuloplasty was performed under general anesthesia and echocardiographic guidance. Pericardial effusion was not observed after the procedure. However, LV function could not be ameliorated and continued to diminish. There was no cardiac activity in the fetus two hours after the intervention. Aortic valvuloplasty *in utero* for AS is technically feasible. Mortality is mainly associated with technical errors, LV function, and the degree of endofibroelastosis in the effected fetuses.

Özet– Aort darlığı olan fetusta mortalite ve morbidite, aort darlığının hemodinamik etkilerinin derecesi, sol ventrikülün (SV) uyumu, gelişimi ve fonksiyonları ile ilişkilidir. Kritik aort darlığı durumunda, rahim içinde ölüm ve hidrops gelişebileceği iyi bilinmektedır. Yirmi üç haftalık bir fetusta kritik aort darlığı ve eşlik eden kardiyomegali, SV'de genişleme ve kontraktilite kaybı, mitral yetersizliği saptandı. Doppler incelemesinde arkus aortada duktus arteriyozis yoluyla ters yönde akım ve duktus venosusta ters a-dalgası vardı. Fetusta asit, kafa derisi, yüz ve ciltte masif ödem ile birlikte hidrops vardı. Fetal amniyosentez normal bulundu. Genel anestezi ve ekokardiyografi kılavuzluğunda aort kapağına valvüloplasti yapıldı. İşlem sonrası ekokardiyografide perikart efüzyonu görülmedi. Ancak SV fonksiyonlarının düzelmediği ve azaldığı gözlendi. İşlemden iki saat sonra fetusta kalp hareketleri durdu. Fetusta aort darlığında uterus içi aort kapağına valvüloplasti uygulaması teknik olarak mümkündür. Mortalite teknik başarı yanında, SV fonksiyonları ve endofibroelastosis derecesine doğrudan bağlıdır.

One of the most severe forms of congenital cardiac malformations is life-threatening, severe critical aortic stenosis (AS). Neonatal mortality and morbidity are associated with left ventricular (LV) size, hypoplasia, dysfunction, aortic valve or arch hypoplasia, endocardial fibroelastosis, and mitral regurgitation. Most forms of the condition are well-tolerated during the fetal period, but in case of critical AS, the development of hydrops and death *in utero* commonly occurs.[1] Even when the obstruction to the LV outflow tract is relieved after birth, the LV often fails to maintain cardiac output. It is hypothesized that early rescue of the ventricles from an unfavorable environment may promote healthier ventricular and vascular growth and improve postnatal outcomes. Hence, fetal cardiac valvuloplasty has been proposed for progressive cardiac disease with a poor prognosis, such as critical AS and pulmonary atresia with an intact ventricular septum, and balloon atrial septostomy has been proposed for hypoplastic left heart syndrome (HLHS) or simple transposition of

Abbreviations:

AS	*Aortic stenosis*
BAV	*Balloon aortic valvuloplasty*
HLHS	*Hypoplastic left heart syndrome*
LV	*Left ventricular*
RV	*Right ventricle*

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the great arteries with closed or restrictive inter-atrial communication.[2-5]

We performed balloon aortic valvuloplasty (BAV) in a fetus with severe critical AS to improve the dismal prognosis for this condition in an attempt to relieve the obstruction of the aortic valve before the development of irreversible LV damage.

CASE REPORT

A 25-year-old pregnant woman at 23-week gestation was referred to the maternal and fetal unit because of a suspicious four-chamber view. Detailed fetal ultrasonography and echocardiography (Voluson 730, 4-8 MHz 4D probe, GE, Austria) revealed visceral and cardiac situs solitus, cardiomegaly (cardiothoracic ratio >0.65) and a dilated LV (LV length: 25 mm; Z value: +2.96) with an end-diastolic diameter of 15 mm (Z value: +4.02). There was LV compression of the right ventricle (RV) (RV length: 15 mm, end-diastolic diameter: 7.0 mm; normal Z value) and very poor contractility of the LV with a hyperechogenic rim suggesting endocardial fibroelastosis (ejection fraction: 24%), mitral regurgitation, and aortic valve stenosis (aortic annulus diameter: 1.5-2 mm; Z value: -9-13) without interruption and signs of coarctation, as well as a prominent and dilated pulmonary artery (5.5 mm; Z value: +20) (Fig. 1) (Video 1*). All reported Z-scores were based on the gestational age and calculated from the Royal Brompton Hospital study (Table 1).[6] The ductal arch was well developed, but there was a reversal of flow in the aortic arch through the ductus arteriosis. The fetus had ascites, massive scalp, face, and skin edema, and a reversed a-wave in the ductus venosus. These findings suggested that the fetus had hydrops and heart failure because of severe critical AS due to decreased left atrial pressure. There was no extracardiac anomaly. Amniocentesis was performed and a fetal karyotype test yielded a normal result. The parents were informed about the poor prognosis and high risk for *in utero* mortality and were advised to consider termination. The parents did not accept a fetal intervention at 23 weeks. Follow-up on the fetal condition was performed weekly, during which progressive deterioration of heart failure was observed. Aortic balloon valvuloplasty or no interventions were discussed at our perinatology-neonatology-pediatric cardiology committee and we decided

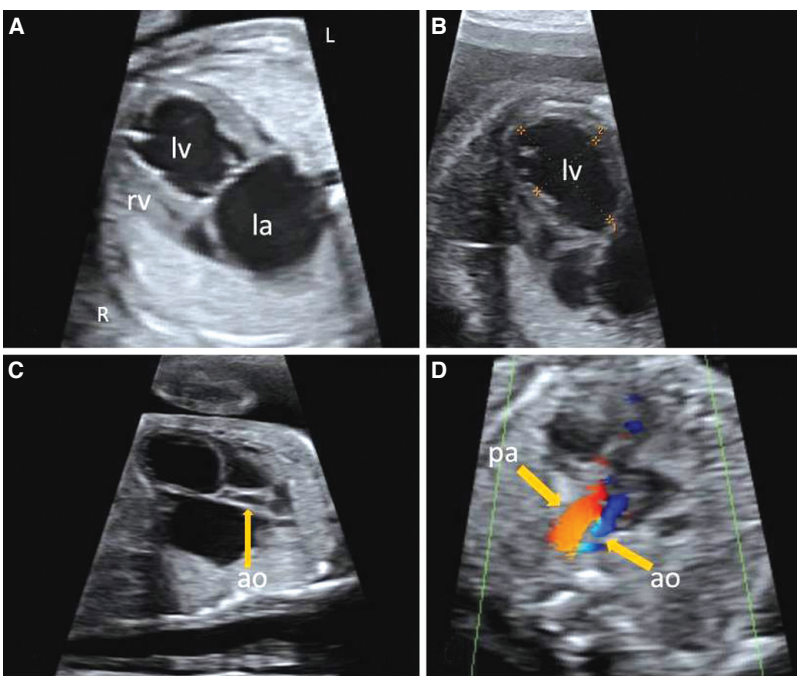

Figure 1. Echocardiographic images: **(A, B)** Dilated left ventricle, left atrium and compressed right ventricle. **(C)** Critical aortic stenosis. **(D)** In the three-vessel and tracheal view, the aortic diameter is much smaller than the pulmonary artery with reversed flow in the aortic arch.

Table 1. Z-score equations

Aortic annulus diameter (cm)
Mean = (0.02415 x GA) - 0.17158
SD = (0.00206 x GA) - 0.00519
Ascending aortic diameter (cm)
Mean = (0.02413 x GA) - 0.12588
SD = (0.00205 x GA) + 0.00587
LV long-axis dimension (cm)
Mean = (0.09541 x GA) - 0.55304
SD = (0.01149 x GA) - 0.06876
LV short -axis dimension (cm)
Mean = (0.05981 x GA) - 0.51997
SD = (0.00784 x GA) - 0.06281
MV annulus diameter (cm)
Mean = (0.03482 x GA) - 0.21035
SD = (0.00222 x GA) + 0.01698
RV long-axis dimension (cm)
Mean = (0.09512 x GA) - 0.68831
SD = (0.00890 x GA) - 0.01642

GA: Gestational age based on dates or first trimester ultrasound if available.

to propose BAV to the parents. At 28 weeks gestation, the mother gave consent for the intervention. After the parents were fully informed about the extremely difficult nature of the procedure and the high risk for death *in utero*, aortic valvuloplasy was attempted.

In the operating room, general anesthesia was administered to avoid maternal movements and to ensure fetal anesthesia. With the fetus in an appropriate position, in which the fetal thorax was near the ultrasound probe and the fetal heart was in the apical position, a throchar was inserted into the LV under ultrasound guidance and passed through the aortic orifice with an 18 G, 15 mm transabdominal chorionic villus needle (Trocar Needle, Cook Medical, Bloomington, IN, USA). A 2.6-Fr standard coronary balloon catheter (3.5 mm / 1.0 cm) was inserted over a guide wire and was positioned across the aortic valve. When the correct balloon position was achieved, the balloon catheter was inflated (Fig. 2) (Video 2*). After demonstration of aortic valve dilatation, the balloon catheter and needle were withdrawn. After the BAV, no pericardial effusion was observed. A short episode of severe fetal bradycardia occurred immediately after puncture of the LV and could not be reversed using atropine. The fetal heart rate remained unstable and bradycardia persisted. There was no further bleeding and no maternal complications. Fetal echocardiography performed one hour after the procedure showed antegrade flow through the aortic valve, but the poor LV function could not be ameliorated. There was no fetal heart activity two hours after the intervention and, subsequently vaginal delivery was induced. The fetus weight 2.500 g and had massive ascites and edema (Fig. 3).

DISCUSSION

Severely critical AS in midtrimester may lead to myocardial damage, resulting in endocardial fibroelastosis and HLHS. The latter is lethal when left untreated, and occurs when the LV cannot support the systemic circulation. In some mid-gestation fetuses with severe AS, the LV size is normal or even enlarged, but as the gestation progresses, left heart growth becomes retarded. Thus, parents face the difficult choice between termination of the pregnancy and having a baby with lifelong cardiac disability. Theoretically, early relief of fetal AS might preserve left heart function and flow *in utero* and perhaps prevent HLHS, giving rise to attempts at fetal aortic valvuloplasty for the past two decades.[1-5,7] The second consequence of AS in the non-adapted LV structure is LV dilation. In this case, endocardial fibroelastosis and myocardial damage occur due to shear stress and LV pressure, respectively. This usually results in fetal mortality because of the increasing severity of heart failure. In the case of AS with LV dilation, progressive LV dysfunction is often

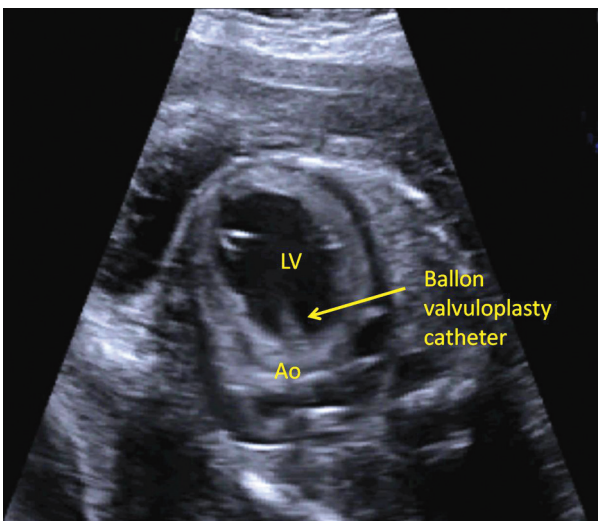

Figure 2. Echocardiograhic image showing introducer cannula and ballon catheter inflation in the aortic valve.

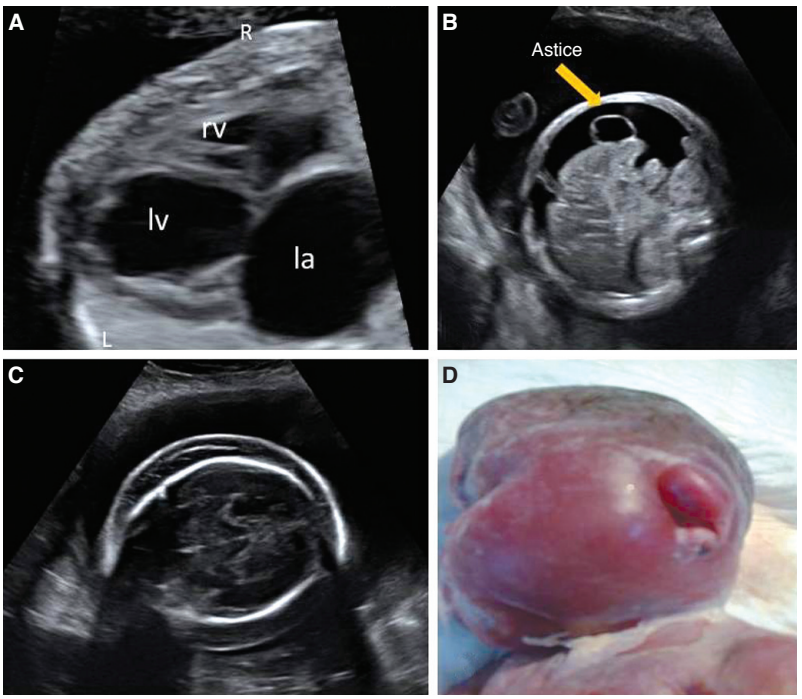

Figure 3. **(A)** Dilated left ventricle and left atrium compared to the right ventricle. **(B)** Ascites. **(C)** Scalp edema. **(D)** Postnatal appearance of the fetus with neck, face and scalp edema.

fatal, leading to heart failure and hydrops fetalis.

Fetal BAV is still a subject of contentious debate and even those with experience in performing the procedure admit that complications are common and that outcomes are extremely variable. It has yet to be demonstrated convincingly that the long-term benefits justify the treatment. The primary goal of BAV is to prevent the development of HLHS or conversely, the development of LV dilatation. It has been shown that fetal aortic valvulopasty is technically feasible in more than 70-74% of cases. Some evidence for improvement in fetal left heart hemodynamics has been demonstrated, sufficient to alter the natural history and to achieve a biventricular circulation in 29% of the affected fetuses,[1,7-9] but the long-term outcome is still not known. This report provides support to previous experience in other centers[2-5] that fetal echocardiography-guided percutaneous BAV *in utero* is technically feasible and can be performed, but we must clarify BAV indications, parents' choice, and the outcomes for the different BAV indications. If parents decide to continue pregnancy, the choice of valvuloplasty should be mentioned and be offered for possible prevention of heart failure progression. In our case, the fetus had hydrops fetalis and severe heart failure with endocardial fibroelastosis. Initially, indecisive parents were indeed an impediment to early intervention. It was not possible to wait to perform a postnatal intervention. Although the intervention resulted in fetal mortality, we believe that it was technically feasible. However, it is clear that some technical modifications are necessary for this procedure to be safe and reliable and to yield a clinically favorable outcome. With promising results and growing experience, there is no doubt that fetal outcomes of *in utero* valvuloplasty will be better in the future.

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****Supplementary video files associated with this article can be found in the online version of the journal.***

REFERENCES

1. Arzt W, Wertaschnigg D, Veit I, Klement F, Gitter R, Tulzer G. Intrauterine aortic valvuloplasty in fetuses with critical aortic stenosis: experience and results of 24 procedures. Ultrasound Obstet Gynecol 2011;37:689-95.
2. Maxwell D, Allan L, Tynan MJ. Balloon dilatation of the aortic valve in the fetus: a report of two cases. Br Heart J

1991;65:256-8.

3. Tworetzky W, Wilkins-Haug L, Jennings RW, van der Velde ME, Marshall AC, Marx GR, et al. Balloon dilation of severe aortic stenosis in the fetus: potential for prevention of hypoplastic left heart syndrome: candidate selection, technique, and results of successful intervention. Circulation 2004;110:2125-31.
4. Marshall AC, van der Velde ME, Tworetzky W, Gomez CA, Wilkins-Haug L, Benson CB, et al. Creation of an atrial septal defect in utero for fetuses with hypoplastic left heart syndrome and intact or highly restrictive atrial septum. Circulation 2004;110:253-8.
5. Lopes LM, Cha SC, Kajita LJ, Aiello VD, Jatene A, Zugaib M. Balloon dilatation of the aortic valve in the fetus. A case report. Fetal Diagn Ther 1996;11:296-300.
6. Schneider C, McCrindle BW, Carvalho JS, Hornberger LK, McCarthy KP, Daubeney PE. Development of Z-scores for fetal cardiac dimensions from echocardiography. Ultrasound Obstet Gynecol 2005;26:599-605.
7. Kohl T, Sharland G, Allan LD, Gembruch U, Chaoui R, Lopes LM, et al. World experience of percutaneous ultrasound-guided balloon valvuloplasty in human fetuses with severe aortic valve obstruction. Am J Cardiol 2000;85:1230-3.
8. Selamet Tierney ES, Wald RM, McElhinney DB, Marshall AC, Benson CB, Colan SD, et al. Changes in left heart hemodynamics after technically successful in-utero aortic valvuloplasty. Ultrasound Obstet Gynecol 2007;30:715-20.
9. McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, et al. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation 2009;120:1482-90.

Key words: Aortic valve/abnormalities; aortic valve stenosis/therapy; balloon valvuloplasty; catheterization/methods; fetus/surgery; gestational age; ultrasonography, prenatal.

Anahtar sözcükler: Aort kapağı/anormallik; aort kapağı darlığı/tedavi; balon valvuloplasti; kateterizasyon/yöntem; fetus/cerrahi; gestasyonel yaş; ultrasonografi, prenatal.