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Aorto-left ventricular Tunnel – Prenatal Diagnosis and Outcome

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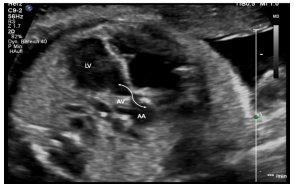
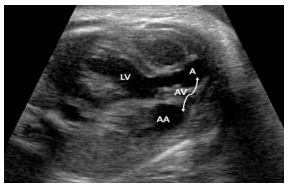
Purpose

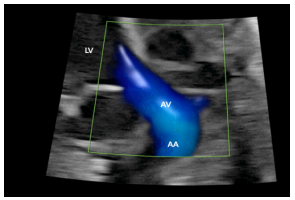
Aorto-left ventricular tunnel (ALVT) is an extremely rare, albeit prenatally detectable, extracardiac channel which connects the ascending aorta to the cavity of the left ventricle.



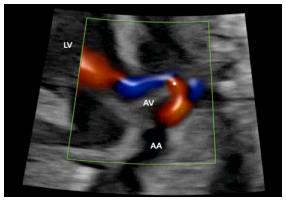
Left ventricular outflow tract view demonstrating a large paravalvular tunnel (arrow) bypassing the atretic aortic valve (AV). The left ventricle (LV) is dilated as well as the ascending Aorta (AA).



Large aneurysm (A) of the left outflow tract and the ascending aorta (AA) in a fetus with aorto-left ventricular tunnel (arrow) and highly stenotic aortic valve (AV).



Color Doppler of the left ventricular outflow tract demonstrating the systolic flow from the left ventricle (LV) over the aortic valve (AV) into the dilated ascending aorta (AA) while the tunnel cannot be seen.



Color Doppler of the left ventricular outflow tract demonstrating the diastolic regurgitation from the ascending aorta (AA) into the left ventricle (LV) over a narrow paravalvular tunnel (asterix) bypassing the aortic valve (AV).

Material and Methods

All ALVT diagnosed prenatally (2006-2022) in five tertiary referral centers were retrospectively assessed for prenatal ultrasound findings, intrauterine course, postnatal outcome and surgical treatment. Focus was on the size of the tunnel and alterations of perfusion of the left ventricular outflow tract and aortic arch.

Results

13 fetuses were diagnosed with ALVT at a mean gestational age of 23.4 weeks. All cases were associated with severe dilatation of the left ventricle and to and fro flow in the left outflow tract. Signs of congestive heart failure were present in five fetuses, four of which were terminated and one died in the neonatal period. One was terminated for a PIK3CA-associated overgrowth syndrome. One fetus died in utero at 34 weeks without prior signs of cardiac failure. Of the five survivors, two underwent Ross procedure. In both cases the prenatal left ventricular outflow was exclusively via a large tunnel. The remaining three neonates underwent patch closure of the tunnel. In these cases the prenatal outflow of the left ventricle was via the aortic valve and simultaneously over the tunnel.

Case	Sex	GA	RD	Prenatal findings	Postnatal/ autopsy findings	Outcome	Follow up
1	m	21	VSD, aortic insufficiency	ALVT right anterior (3.6mm at 21 weeks), antegrade flow over AV, normal flow in AoA, AA aneurysm, CHF, PE	Autopsy: RVOT obstruction	TOP at 21.6 weeks	
2	m	18	ToF	ALVT right anterior (3mm at 18 weeks), antegrade flow over AV, to and fro flow in AoA, AA aneurysm, CHF, PE, HF	Autopsy: AV dysplasia with stenosis	TOP at 20.1 weeks	
3	N/a	18	Cardiomegaly, VSD, aortic stenosis	ALVT right anterior (2.5mm at 18 weeks), antegrade flow over AV, to and fro flow in AoA, AA aneurysm, CHF, PE, pleural effusion	Autopsy: AV dysplasia with stenosis, non compacted LV myocardium	TOP at 19.5 weeks	
4	f	21	Right aortic arch	ALVT right anterior (3.7mm at 21 weeks) antegrade flow over AV, to and fro flow in AoA, AA aneurysm, CHF, PE		TOP at 21.5 weeks	
5	m	22	VSD, dilated LV, RV hypertrophy	ALVT right anterior (2mm at 22 weeks, 3mm at 28 weeks, 6mm at 30 weeks), antegrade flow over AV, antegrade flow AoA, AA aneurysm	Hexadactyly, PIK3CA-associated overgrowth syndrome	TOP at 33.1 weeks	
6	m	24	AAVS	ALVT left anterior (4mm at 30 weeks), antegrade flow over AV, to and fro flow in AoA, LOT aneurysm, AA aneurysm	Autopsy: hypoplastic RCA, chronic ischemic myocardial lesion	IUFD 34 weeks	
7	m	31	ToF	ALVT right anterior (6mm at 36 weeks), antegrade flow over AV, to and fro flow in AoA, AA aneurysm, CHF, PE, HF	Dysplastic MV, non compacted LV myocardium, severe CHF, pulmonary edema, hepatomegaly	Neonatal death	
8	f	20	ToF	ALVT right anterior (4mm at 30 weeks), no flow over AV, antegrade flow in AoA, AA aneurysm	Aortic atresia, ASD II	Neonatal Ross procedure	Tolerable conduit insufficiency, 6 years old
9	m	30	ToF	ALVT right anterior (6mm at 30 weeks), no flow over AV, to and fro flow in AoA, LOT aneurysm, AA aneurysm	Severe AS, non compacted LV myocardium, ASD II	Neonatal patch closure of ALVT, commissurotomy of aortic valve	Secondary Ross operation at age of 3 months, mild conduit insufficiency, 7 years old
10	m	25	ToF	ALVT right anterior (2mm at 30 weeks), severe PS, antegrade flow over AV, antegrade flow in AoA, LOT aneurysm	Origin of RCA in ALVT, normal LCA origin	Neonatal patch closure of ALVT, creation of RCA tunnel, valvotomy and commissurotomy of PV	Mild residual PI, 7 months old
11	m	27	Coronary fistula	ALVT right anterior (2mm at 30 weeks), antegrade flow over AV, antegrade flow in AoA		Neonatal patch closure of ALVT	Thriving, 3 years old
12	m	38	VSD, aortic insufficiency	ALVT right anterior (5mm at 38 weeks), antegrade flow over AV, to and fro flow flow in AoA, AA aneurysm	Origin of RCA in ALVT, normal LCA origin, dysplastic AV, aortic stenosis and insufficiency	Neonatal patch closure of ALVT	Moderate AS and AI, AA aneurysm, Bentall procedure (at age 13 years), 15 years old
13	f	23	TA Type I, VSD	ALVT right anterior (1.5mm at 23 weeks, 2mm at 35 weeks)		Ongoing pregnancy	

AA, ascending aorta; AAVS, absent aortic valve syndrome; AI, aortic insufficiency; ALVT, aorto-left ventricular tunnel; AoA, aortic arch; AS, aortic stenosis; ASD II, secundum atrial septal defect; AV, aortic valve; CHF, congestive heart failure; HF, hydrops fetalis; IUFD, intrauterine fetal death; LCA, left coronary artery; LOT, left outflow tract; LV, left ventricle; MV, mitral valve; N/a, not applicable; PE, pericardial effusion, PI, pulmonary insufficiency; PS, pulmonary stenosis; PV, pulmonary valve; RCA, right coronary artery; RD, referral diagnosis; RVOT, right ventricular outflow tract; TA, truncus arteriosus communis; ToF, tetralogy of Fallot; TOP, termination of pregnancy; VSD, ventricular septal defect

Conclusion

Prenatal diagnosis of ALVT should be considered in the presence of left ventricular hypertrophy, dilatation of the aortic root and to and fro flow in the aortic outflow tract. Signs of heart failure are associated with unfavorable outcome. Large tunnels, particularly in combination with absence of flow over the aortic valve, may be an unfavorable predictor of surgical repair.

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