Acquired Right Ventricle Outflow Tract Obstruction in Twin-Twin Transfusion Syndrome

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Introduction

Twin-Twin Transfusion Syndrome (TTTS) complicates 4 to 26% of diamniotic monochorionic twin gestations. The majority of deaths associated with TTTS are due to cardiovascular compromise from congestive heart failure. Right ventricle outflow tract obstruction (RVOTO) may occur in the recipient twin in at least 9% of pregnancies complicated by TTTS. The etiology is not completely understood. RVOTO can potentially progress in utero and may worsen neonatal outcome.

Methods

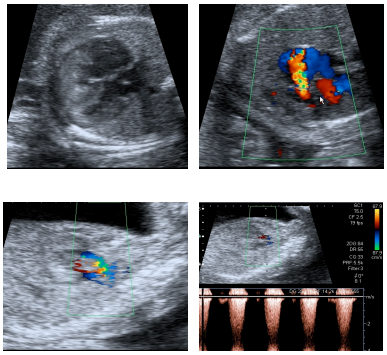
Retrospective review of all cases of TTTS treated at our hospital between March 2008- November 2010.
Demographic data, fetal echocardiograms, perinatal and postnatal outcome and catheterization records were reviewed.

Results

Fifty-two pregnancies complicated by TTTS were identified. 6 recipient twins (11,53%) had RVOTO prenatal diagnosis. None of the donor twins had structural heart disease, and no major additional structural heart disease was identified in the recipient twins.
Gestational age range was 16-23 weeks at RVOTO diagnosis. 5/6 cases received photocoagulation (83%).

Echocardiographic data at diagnosis

Case	TV	RV	PV	LV	Duct
1	Severe TR grad 100 mmHg	Hypertrophic	Dysplasic grad 36 mmHg	Normal	Retrograde
2	Severe TR grad 80 mmHg	Hypertrophic	Dysplasic grad 45 mmHg	Normal	Retrograde
3	Normal	Normal	Grad 36 mmHg	Normal	Retrograde
4	Severe TR grad 43 mmHg	Hypertrophic	Dysplasic grad 23 mmHg	Normal	Retrograde
5	Moderate TR	Hypertrophic	Dysplasic grad 36 mmHg	Hypertrophic	Retrograde
6	Moderate TR	Hypertrophic	Dysplasic grad 40 mmHg	Hypertrophic	Retrograde



Prenatal outcome:

3 of 6 cases (50%) progressed to more severe obstruction during gestation. One case developed progressive stenosis with pulmonary artery calcification. Delivery was between 29-38 weeks and none was indicated for RVOTO. Weight was between 1380 to 2340 gr.

Postnatal outcome:

Case	Echocardiography	PGs	Valvuloplasty	Outcome
1	Severe pulmonary stenosis	Yes	24 h.Infundibular gradient	Stable with propranolol. Dysplasic TV
2	Critical pulmonary stenosis with infundibular stenosis	Yes	3 d. Infundibular gradient	Infundibular resection+ Comisurotomy. Dysplasic TV
3	Moderate pulmonary stenosis	No	75 d. Residual gradient 25 mmHg	Stable. Dysplasic TV
4	Moderate pulmonary stenosis	Yes	17 d. Gradient 12 mmHg	Stable. Dysplasic TV
5	Severe pulmonary stenosis valvular and supravalvular with calcification	Yes	13 d. Gradient 10-15 mmHg	Surgery of supravalvular calcification at 2 m
6	Severe pulmonary stenosis	Yes	3 d. Gradient 5 mmHg	Stable

Pericardial tamponade requiring surgical drainage complicated one procedure.

Conclusions

RVOTO complicates 11,5% of TTTS pregnancies. Diagnosis and possibility of progression does not differ from structural RVOTO. None of our cases suffered fetal hemodynamic compromise and no neonatal deaths occurred despite all patients needing neonatal catheterization. The high incidence of duct dependency underlines the importance of prenatal diagnosis and highlights the indication of echocardiography evaluation in all pregnancies complicated by TTTS.