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# Critical Coarctation of the Aorta in the Donor twin in Twin-twin Transfusion Syndrome and successful treatment with Balloon Angioplasty after Birth: A Case Report

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**Abstract**

Coarctation of aorta (CoA) is a common cause of congenital heart disease (CHD) which is caused by narrowing of aorta. Monochorionic (MC) twins are at increased risk of CHDs, especially acquired CHDs in twin-twin transfusion syndrome (TTTS). CoA is a rare coincidence with TTTS. MC twin pregnancies have been increasing in last decades due to an increase in maternal age and extensive use of assisted reproductive technologies. Multiple cardiac abnormalities in MC twins with TTTS are to be expected due to cardiac hemodynamic changes. Therefore, more attention to this group of patients should be given regarding heart abnormalities. Prenatal diagnosis of CoA is difficult but it is necessary given the importance of treatment after birth. Here we present a case of CoA coexisting in the donor infant of a twin with TTTS.

**Keywords:** Congenital heart disease • Cardiac dysfunction • Coarctation • Aortic stenosis

## Introduction

Twin to twin transfusion syndrome (TTTS) is a rare and serious condition that occurs in monochorionic (MC) pregnancies. It has a high perinatal morbidity and mortality if left untreated [1]. In the recipient fetus biventricular hypertrophy, atrio-ventricular valves regurgitation, cardiomegaly, cardiac dysfunction and finally fetal hydrops may occur all of them mainly due to hypervolemic status. Moreover, right ventricular outflow obstruction (RVOTO), valvar pulmonary stenosis (PS) and atresia and left ventricular hypertrophic cardiomyopathy may also be presented. Donor twin on the other hand, usually shows little cardiac pathologies on echocardiography [2-5]. Different types of cardiac abnormalities in TTTS can be explained by the altered fetal hemodynamics [6]. Fetoscopic laser photocoagulation (FLP) at 16-26 weeks of gestation is the treatment of choice for TTTS. There are evidences of normalization of cardiac function in fetuses with severe TTTS after FLP Acquired structural cardiac anomalies are detected in a proportion of treated pregnancies, as a consequence of the altered hemodynamics [7]. Due to increased rate of CHDs in TTTS, especially PS in recipient twin, prenatal and postnatal echocardiographic assessment is needed [8].

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## Case Presentation

A 27-year-old pregnant woman, primigravida, MC twin pregnancy, with a gestational age of 21 weeks was referred to our center due to stage 3 TTTS. Her pregnancy was a product of in vitro fertilization (IVF). She had no significant past medical history and she did not use any drugs. Fetal echocardiography showed severe coarctation of aorta (CoA) in the descending aorta with retrograde flow in the donor twin (Figure 1). No other major anomalies were detected in the fetuses. FLP was performed at our center for treating TTTS. Accordingly, further development of TTTS was stopped and at 33 weeks of gestation, the pregnancy was terminated by cesarean section due to the onset of labor pains. Both babies were born with good APGAR score and appropriate weight. Postnatal echocardiography of donor twin in the supra-sternal view showed severe narrowing of proximal part of aorta with flow acceleration (Figure 2). In Doppler assessment, we detected a typical saw tooth appearance in the location of coarctation (Figure 3) and apical four chamber view showed

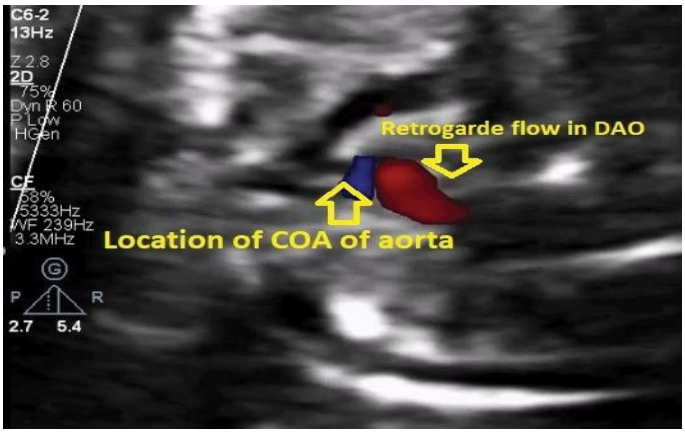


**Figure 1.** Sagittal view shows retrograde (reversal) flow in descendingaorta (DAO), indicating severe coarctation of aorta (COA).

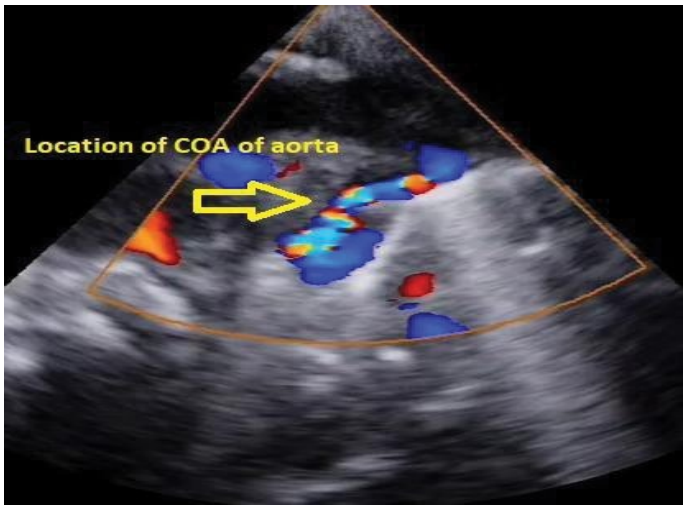


**Figure 2.** Suprasternal view of transthoracic echocardiography shows severe narrowing of proximal portion of aorta (coarctation) with flow acceleration after birth.

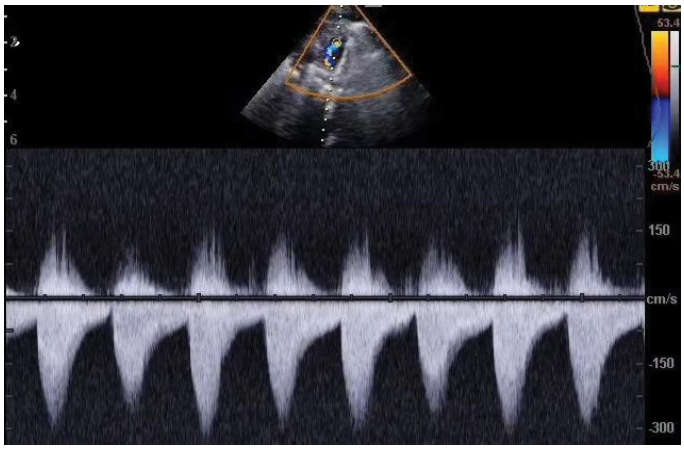


**Figure 3.** Doppler spectral in the location of coarctation shows atypical saw tooth appearance.

left ventricular hypertrophy (LVH), significant left ventricular enlargement (LVE) and left atrial enlargement (LAE) (Figure 4). With a diagnosis of persistent aortic coarctation (Figure 5), donor twin underwent balloon inflation of aorta in the stenotic portion at the age of one month. Balloon inflation of aorta in the stenotic location caused the narrowing to completely resolve (Figures 6-8) and transthoracic echocardiography after balloon angioplasty in suprasternal view showed significant reduction in aortic stenosis (Figure 9).

## Discussion

CoA is a common congenital heart defect that is characterized by a narrowing distal to the aortic arch and the prenatal diagnosis is challenging and difficult [9]. Its coincidence with TTTS is rare and there have been only a few cases in the literature [6]. Assessment of ventricular size is helpful in prenatal diagnosis and the left ventricle is usually smaller than the right ventricle. Also, comparison of the width of the aorta, ductus arteriosus and coarctation shelf in color doppler studies may suggest that the defect will require an intervention after birth [10]. TTTS is associated with an increased prevalence of congenital heart diseases (CHDs). In MC pregnancies with TTTS, the risk of at least one of the infants with CHD is threefold compared to uncomplicated MC twin pregnancies [11]. The development of acquired CHDs in MC twins is associated with TTTS, indicating an influence of hemodynamic alterations on cardiac development. MC twins with TTTS are associated with increased risk of ventricular septal defect (VSD), RVOTO, atrial septal defect (ASD), CoA, and aortic stenosis (AS). Some of them are uncommon, such as CoA and AS [8].

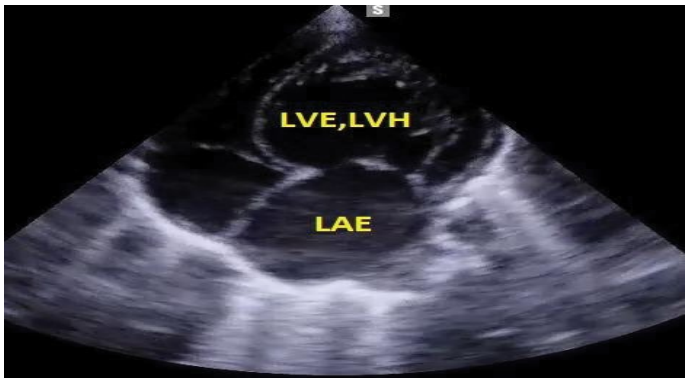


**Figure 4.** Apical four chamber view shows left ventricular hypertrophy (LVH), significant left ventricular enlargement (LVE), and left atrial enlargement (LAE).

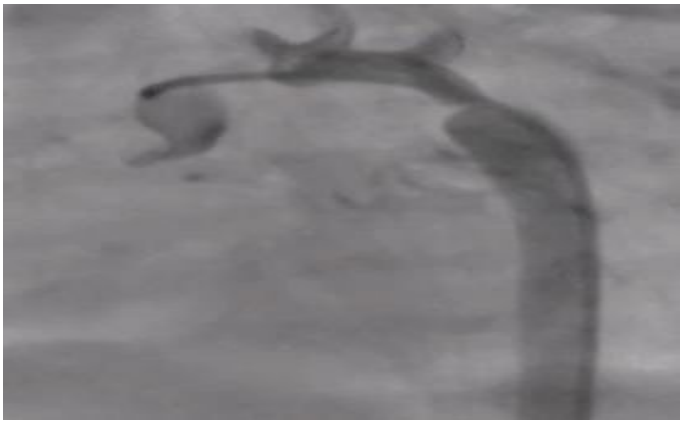

**Figure 5.** Angiographic coronal view of aorta shows significant narrowing of location of coarctation with post stenotic dilation.

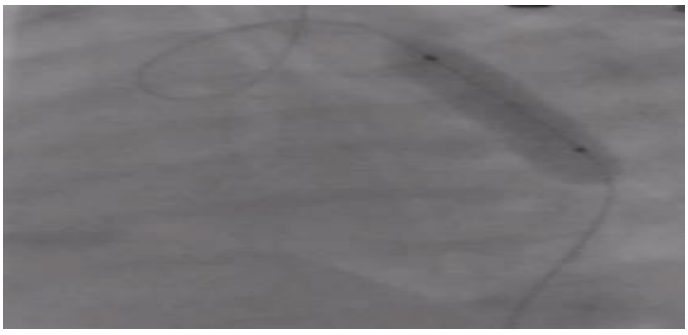

**Figure 6.** Balloon inflation of aorta in stenotic location shows a full dilation of narrowing.

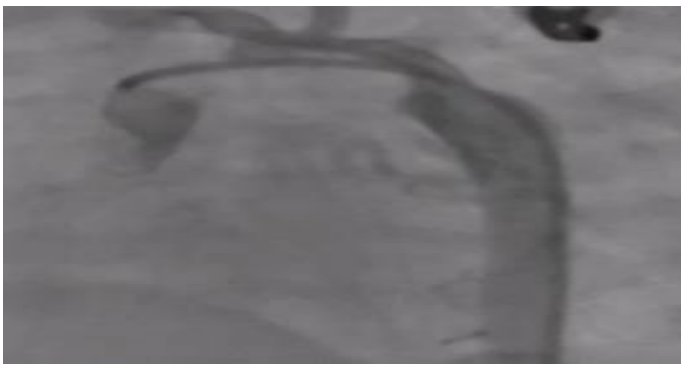

**Figure 7.** Contrast injection in angiography shows significant reduction of stenosis in the location of aorta.

There are reports of a small number of CoA in recipient and donor twins in TTTS. After birth, due to ductal closure, a variety of hemodynamic changes may occur according to the severity of obstruction and critical coarctation increases

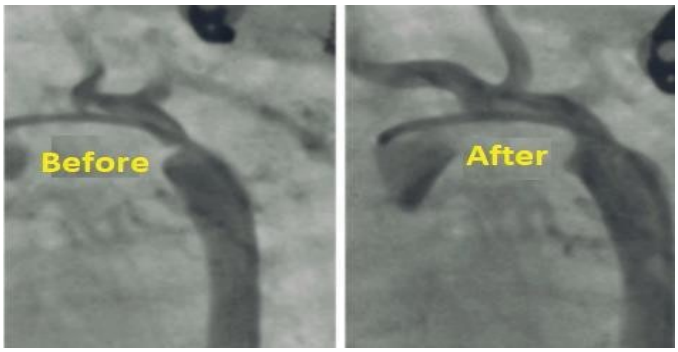


**Figure 8.** Comparison of stenotic portion of aorta before and after balloonangioplasty.

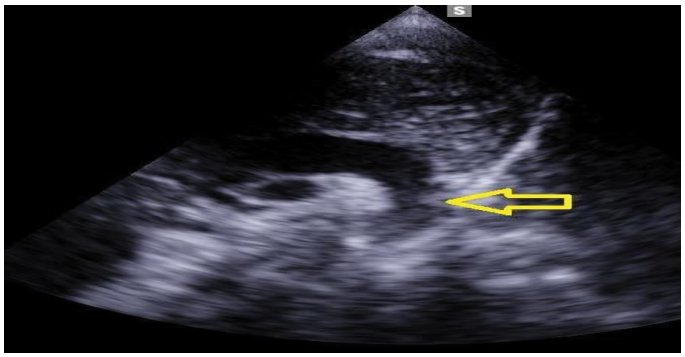

**Figure 9.** Transthoracic echocardiography after balloon angioplasty in asuprasternal view shows significant reduction of stenosis in aorta.

the risk of heart failure and death in infants when the ductus arteriosus closes. So, prenatal diagnosis is important to reduce postnatal complications.

After birth, prostaglandins type 1 is necessary to avoid closure of ductus arteriosus and definite treatment is end-to-end anastomosis, but transcatheter treatment is also an accepted alternative with comparable results [9]. Balloon angioplasty is an acceptable technique, similar to what was done in our case. It is a safe procedure in infants between one and six months of age and also in critically ill patients regardless of age [12]. The best time for CoA correction is in infancy or childhood in order to prevent persistence or late recurrence of systemic hypertension [13].

## Conclusion

It is necessary to pay attention to cardiac abnormalities in MC twins, especially in cases with TTTS, and follow up even if they are treated with FLP during pregnancy and after birth to best treatment.

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