# A Rare Case of Dextrocardia with Situs Inversus Totalis in a Patient of Diabetic Mother

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## ABSTRACT

Infants of diabetic mothers are significantly at higher risk for major congenital malformations, with cardiovascular anomalies that is the most frequent. In this study, we presented a rare case of dextrocardia and situs inversus totalis (mirror-image dextrocardia) with multiple congenital heart anomalies who was born from a diabetic mother.

## 1. Introduction

Although the causes of most birth defects are poorly understood, maternal diabetes is considered as an important risk factor for congenital heart disease (1) . Infants of diabetic mothers are significantly at higher risk for major congenital malformations, cardiovascular anomalies as the most frequent anomaly (2) . Transposition of the great arteries (TGA) , ventricular septal defects (VSD) , double outlet right ventricle, tricuspid atresia, truncus arteriosus and patent ductus arteriosus are the most common types of cardiac malformations that have been seen in infants of diabetic mothers (3, 4) . However, to our knowledge, multiple congenital heart anomalies in conjunction with dextrocardia and situs inversus totalis in infants of diabetic mother have rarely been described in the literature.

Here, we presented a rare case of dextrocardia with situs inversus totalis (mirror-image dextrocardia) with multiple congenital heart anomalies who was born from a diabetic mother and underwent cardiac catheterization in our center.

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## Case Presentation

An 8-years-old girl was presented to the Heshmat hospital, a referral center for congenital heart diseases in north of Iran, with dyspnea, cyanosis, and clubbing. She was a known case of dextrocardia with transposition of the great arteries (TGA) and underwent a palliative shunt 7 years ago. Her mother was diagnosed to have diabetes in the sixth week of pregnancy. She did not started insulin therapy at that time, probably due to low socioeconomic status and poor treatment compliance or other causes. Finally, she was forced by her physician at fourth month of pregnancy to initiate the treatment by insulin. She continued the treatment until the end of pregnancy when her blood glucose returned to the normal limits but did not use it during breast feeding. However, 1.5 years later, she developed diabetes and started using metformin 500 mg three times a day. Later, she started insulin therapy because of her poor glycemic control. The mother was not smoker but his father was a smoker with a 25 pack/year history. Upon admission, the patient had respiratory distress with respiration rate of 35 per minutes. She had central cyanosis and her O2 saturation was 74 percent by pulse oximetery. Palpation at precordium revealed a ventricular tap at lower right sternal border and apex beat at 5th intercostal space in right midclavicular. There was a systolic thrill in right sternal border (RSB) . In auscultation, S1 was normal and S2 was single. A 4/6 holosystolic murmur was heard in RSB.

Chest radiography demonstrated dextrocardia, with base-apex axis located in the right side, cardiomegaly (cardiothoracic ratio: 0.75) , a right aortic arch, and clear lung fields with decreased pulmonary vascular markings. A gastric air bubble was in the right side, and liver shadow was seen in the left side.

The electrocardiogram showed sinus tachycardia, normal QRS axis, inverted P wave axis because of the atrials inversion. There was evidence of associated right and left ventricular hypertrophy because of transposition physiology and pulmonary stenosis. Abdominal ultrasound demonstrated the stomach and spleen on the right side and the liver on the left. The inferior vena cava was in left side.

Echocardiography demonstrated dextrocardia, complete atrial and ventricular situs inversus, severe pulmonary stenosis (PG=92mmHg) , complete transposi-

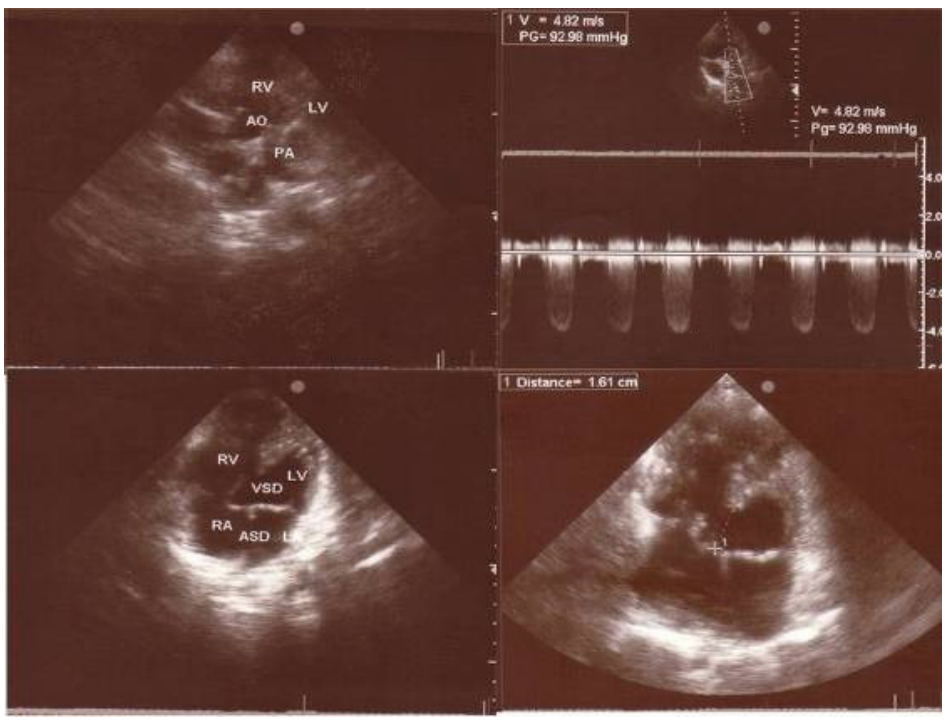


**Figure 1.** Echocardiogms from presented case with dextrocardia and situs inversus totalis. A) Parasternal long axis view; B) Continues wave Doppler of pulmonary artery; C & D) Apical four-chamber view shows ventricular and atrial septal defects. Apical view was taken with cursor probe direction at 9 o'clock position.

RA: right atrium; LA: left atrium; RV: right ventricle; LV: left ventricle; VSD: ventricular septal defect; ASD: atrial septal defect; PA: Pulmonary artery.

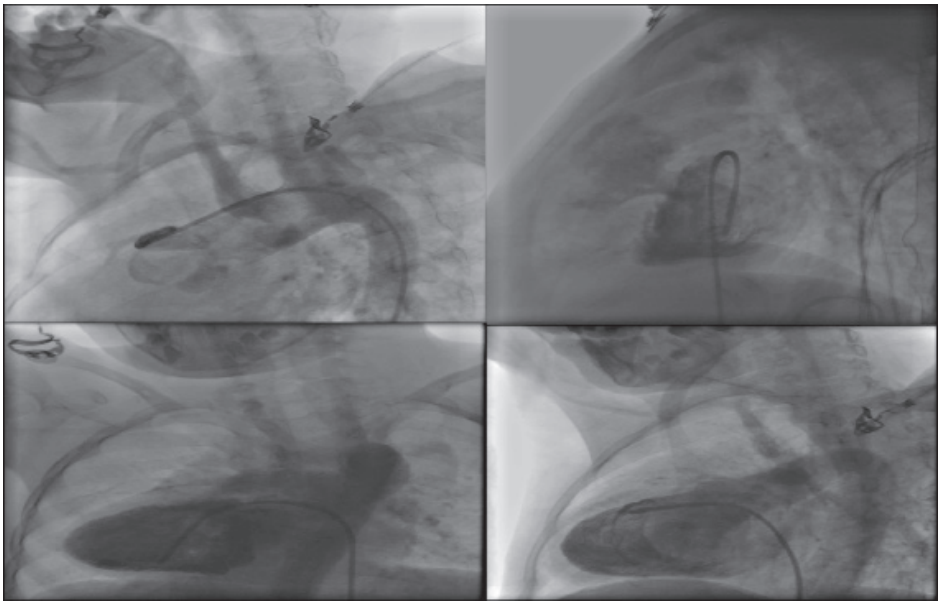


**Figure 2.** Angiograms from presented case with dextrocardia and situs inversus totalis. A) Aortic injection shows normal coronary arteries; B) Right ventricular injection shows ventricular septal defect; C & D) Left ventricular injection shows pulmonary artery stenosis.

tion of the great arteries atrioventricular concordance, and ventriculo-arterial discordance. There was bidirectional shunting through a large-size (16 mm) nonrestrictive perimembranous ventricular septal defect, and there was bilateral shunting through large atrial septal defect (ASD) . Continuous left-to-right shunting due to a large patent ductus arteriosus was also present. In addition, a 15 mm ASD was present and the aortic arch was on the right side (Figure 1) .

Cardiac catheterization was performed and vein catheter was placed through femoral venous and entered inferior vena cava (IVC) and right ventricle in left side position. Systemic sample was desaturated. Right ventricle pressure was systemic. Left ventricle anterior negotiated from right ventricle via VSD. Right ventricle posterior angiogram in anteroposterior (AP) and lateral views showed large VSD, Right Aortic Arch and Dextrocardia. Left ventricle anterior angiogram in AP, left anterior oblique angiogram showed large VSD, severe pulmonary stenosis, TGA and dextrocardia.

Aorta angiogram in LAO and RAO view showed normal coronary artery (NCA) with occluded brachiocephalic-right pulmonary artery (RPA) shunt (Figure 2) .

## Surgical Procedure

Patient who referred to a specialized centre for surgical procedure was underwent modified Fontan operation at that center.

## 2.Discussion

This is a rare case of dextrocardia and situs inversus totalis in combination with multiple congenital heart anomalies who was borne from a diabetic mother. She successfully was underwent cardiac catheterization and surgical operation (modified Fontan) .

Situs inversus totalis is a rare congenital anomaly, with an incidence of 1/10,000 (5) , in which the cardiac apex is located on the right side of the thoracic cavity and the major visceral organs are reversed from their normal positions (6) . Dextrocardia with situs inversus can be associated with congenital heart disease, which is observed in 5-10% of all cases (7) . Common congenital heart defects that have been reported including simple D-transposition of the great arteries which is commonly associated with ventricular septal defect, atrial septal defect, arch obstruction, and atrioventricular valve abnormalities (8) .

Infants of diabetic mother are at increased risk of congenital anomalies, mainly cardiac malformation. Congenital anomalies in infants of diabetic mother is thought to be correlated with teratogenic effects of high blood sugar

during the periconception and embryogenesis period (1, 2) . The association of dextrocardia and situs inversus with diabetic mother is rarely reported in the literature. In our case, poor glycemic control in the first trimester of pregnancy is probably the main reason for congenital heart defects.

Coronary angiography was first reported in dextrocardia in 1974 (9) . The most important modifications in performing coronary angiography in such patients are opposite-direction catheter rotations and mirror-image angiographic angles, i.e. anticlockwise rotation needed in the ascending aorta for right coronary artery and reversing the required right anterior oblique RAO/LAO angles, keeping the cranial/caudal tilts the same (10) .

In conclusion, multiple cardiac defects including dextrocardia combined with situs inversus totalis and transposition of the great arteries could be a rare complication of uncontrolled diabetes during pregnancy. All pregnant women except low risk ones should be routinely screened for high blood sugar in the preconception period. Moreover, it seems that preconception care of diabetes using standard dietary regimen, insulin therapy and self-monitoring of blood glucose, can reduce the risk of congenital anomaly in infants of diabetic mothers (11) . In addition, cardiac catheterization and angiography could be successfully performed to evaluate congenital heart defects, assess valves functions and calculate the venous pressure plus oxygen saturation using standard methods.

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