Truncus arteriosus Type ii

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خلفية الدراسة:
ان الجذع الشرياني يعتبر من اللحالات الغير الشائعة من حالات تشوهات القلب الخلقية ويتميز هذا التشوه بأن الشريان الرئوي والابهر يخرجان من جذع شرياني واحد ينشأ من البطينين.

تقديم حالة
راجعت مستشفى بابل للولادة والاطفال طفلة تبلغ من العمر ثلاثة اشهر تعاني من ازرقاق خفيف وصعوبة في التنفس والرضاعة .كانت هذه الطفلة قد راجعت مستشفيين وشخصت بتلك المستشفيين على انها تعاني من فتحة بين البطينين وارتفاع في الضغط الرئوي واعطيت العلاج ولكن لم تتحسن الحالة
لذا راجعت مستشفى بابل للولادة والاطفال وشخصت حالة الطفلة بأنها نوع من الجذع الشرياني :النوع الثاني واعطيت علاج لعجز القلب وارتفاع الضغط الرئوي وتحسنت حالة الطفلة وبعد استقرار وضع الطفلة احيلت الى مركز تخصصي في تركيا لعدم وجود مثل هذا المركز في العراق واجريت لها التدخلات الجراحية وهي دون الشهر السادس من العمر .

الاستنتاجات
ان حالة الجذع الشرياني يجب ان تأخذ بنظر الاعتبار عند تقييم أي رضيع يعاني من ازرقاق خفيف وعجز القلب ويمكن تشخيص الحالة بواسطة الايكو وان تتم التداخلات الجراحية قبل الشهر السادس من العمر لمنع مضاعفات الاوعية الدموية الرئوية .

ABSTRACT

Background : Truncus arteriosus (TA) is an uncommon congenital cardiovascular anomaly that is characterized by a single arterial trunk arising from the normally formed ventricles by means of a single semilunar valve (ie, truncal valve).

Case presentation : A 3-month-old female infant product of spontaneous vaginal delivery complaining from dyspnea , tachypnea , mild cyanosis since birth , decrease in feeding and activity ,She was admitted twice in different hospitals and diagnose as a case of VSD with pulmonary hypertention , given treatment but no response , the family picked her to Babylon for maternity and children hospital . ECHO and cardiac CT- angiography was done were revealed truncus arteriosus.
She was treated by anti failure and drug which lower pulmonary vascular resistance, her condition improve and become stable . after that they went to turkey where operation was done in specialized cardiac centre before age of six month.

Conclusion : In any infant with signs and symptoms of congestive heart failure and mild cyanosis truncus arteriosus shoud be suspected.
Corrective operation should be done in first six month of age

Truncus arteriosus (TA) is a rare cardiac disorder,[1]account of about (0.7%-1.4%) of all congenital heart disease, in which aorta and pulmonary artery were not separated completely during fetal development and both originate jointly from left ventricle[2]. It has an incidence of about 0.3/10,000 live birth,no striking sex difference although most series contain male more than female.[3,4].

Etiology is multifactorial and it may be associated with microdeletion chromosome 22q11.2, maternal diabetes has been implicated as a risk factor for truncus arteriosus.[5]

The number of truncal valve cusps varies from 2 to as many as 6 and the valve may stenotic,regurgitant, or both[6]. Its had been classified by Collet and Edwards in 4 different types in 1949.

Type I in which the pulmonary arteries can arise together from the posterior left side of the persistent truncus arteriosus and then divided into left and right pulmonary

arteries, in type II and III, no main pulmonary artery is present, and the right and left pulmonary arteries arise from separated orifice on posterior(type II) or lateral (type III) aspect of truncus arteriosus. Type IV truncus is a term
no longer used, this essentially a form of pulmonary atresia.(6)
During past 20 years, diagnostic and therapeutic methods along with immediate intervention for surgery in infancy with complete reparation of VSD lead to evident improvement and less morbidity in newborns. Truncus arteriosus canbe diagnosed in utero by fetal echocardiography.(7)

Case presentation: A 3- month- old female infant, product of normal vaginal delivary ,in a consanguineous marriage, she was admitted because of repeated chest infection ,difficulty in breathing and feeding with mild cyanosis since birth, ECHO(echo cardio graphy) was done and was diagnosed as ventricular sepal defect(VSD) and pulmonary hypertension, treatment was given but without benefit and patient still complaining , then they consult another cardiologist with the same result, so they came to our hospital (Babylon for maternity and children hospital) .
On examination. The Patient had mildly cyanosis, dyspneoic, tachypnic, Pulse rate=120 beat/min, bounding peripheral pulse.
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Respiratory rate =62breath/min,SPO2=90% at room air, no dysmorphic feature, Weight=3.8Kg, length=53 cm,
Cardiovascular system : normal S1, loud S2 and single ,on right side loud pan systolic murmur maximal at lower right border radiate to entire pericardium, grade 4/6 systolic thrill ,apex beat on right side at 5th intercostals space ,chest bilateral fine crepitation.
Abdomin : Soft with mild hepatomegaly
Work up: complete blood picture; all indices are normal , serum electrolytes were normal, Liver &renal function tests, plasma glucose all were normal.
CXR(Chest X ray) : Dextrocardia , cardiomegaly . (plethoric lung) as shown in figure (1).

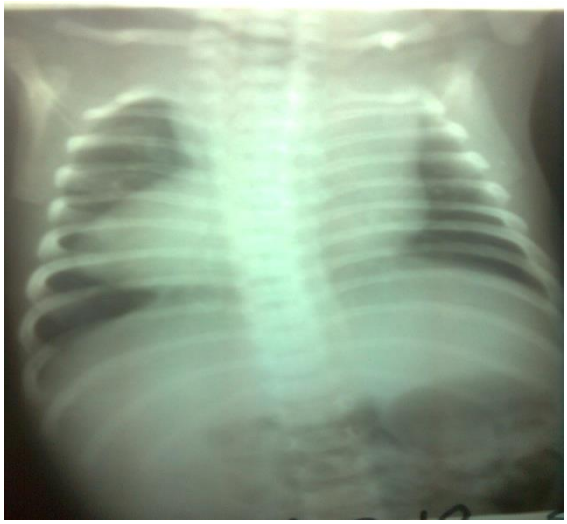

Figure (1):CXR ,A-P View

ECG**(** Electerocardigraphy) :sinus tachycardia, right axis deviation, biventricular hypertrophy. ECHO :situs solitus, dextrocardia , biventricular hypertrophy(RVH), (LVH), dilated right atrium(RA). Large atrial septal defect (ASD) secondum, Large subtruncal ventricle septal defect(VSD) with left to right shunt. Single trunk with mild incompetence guadrivalent valve with both pulmonary arteries arise from

posterior aspect of trunk (truncus arteriosus type II) as shown in figure 2 .

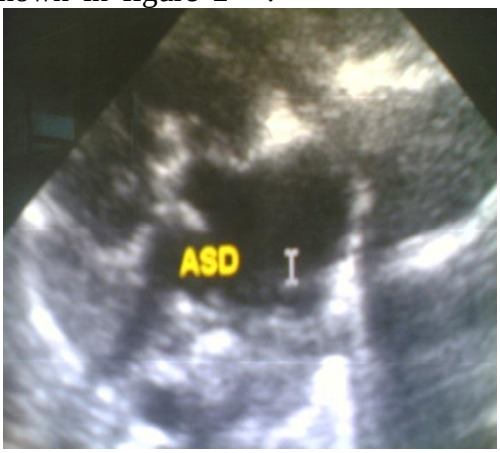

Figure 2 A

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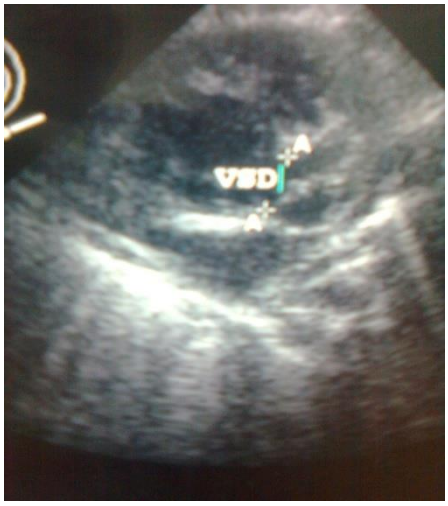

Figure 2 B

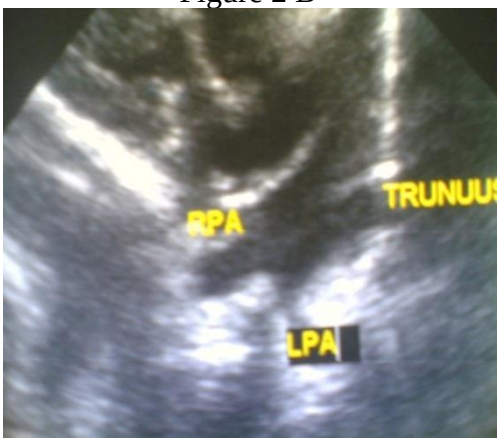

Figure 2 C

Cardiac CT-angiography: Reveal dextrocardia, large ASD secondum, large subtruncal VSD, single trunk arise from both ventricles,absence of pulmonary artery from right ventricle,both pulmonary arteries arise from posterior aspect of the trunk , as shown in figure (3) .

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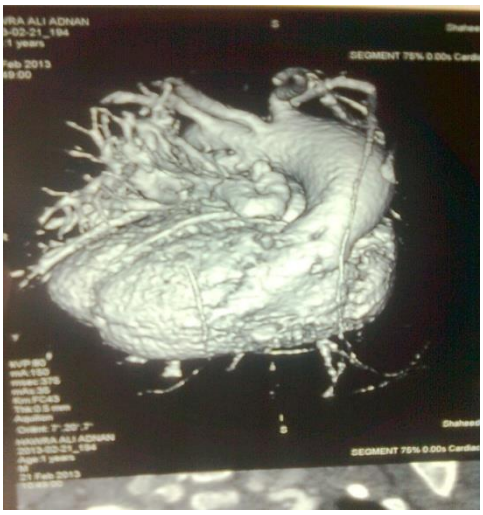

Figure(3):cardiac CT-angiography

So the diagnosis was truncus arteriosus type II and patient received antifailure treatment. Total surgical correction was done at age of six month in turkey.

Literature review : Persistent truncus arteriosus is an uncommon congenital cardiovascular malformation. Truncus arteriosus usually occurs as an isolated cardiovascular malformation, although on occasion it has been reported in association with anomalies of other systems, particularly the DiGeorge or velocardiofacial syndrome (microdeletion chromosome 22q11.2) [8,9].During the last 25 years, surgical correction of truncus arteriosus during infancy has become routine [10]. Among 400 cases of truncus arteriosus from four publications reviewed by Fuglestad et al. [11], the truncal valve was tricuspid in 277 (69%), quadricuspid in 86 (22%), bicuspid in 35 (9%), pentacuspid in 1 (0.3%), and unicommissural in 1 (0.3%). The semilunar valve is in fibrous continuity with the mitral valve in all patients but is continuous with the tricuspid valve in only a minority.

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Type I truncus arteriosus is observed in 48% to 68% of patients, type II in 29% to 48% ,and type III in 6% to 10% [6]. In type II, the left pulmonary artery ostium is generally somewhat higher than that of the right pulmonary artery. Rarely, in the setting of interrupted aortic arch, this ostium may arise to the right of the right pulmonary artery ostium and cause crossing of the pulmonary arteries posterior to the truncus arteriosus [12]. Of the Mayo Clinic's previously published series of patients with truncus arteriosus, 16% (11 of 70) had only a single pulmonary artery [13]. In 9 of the 11 patients, the pulmonary artery was absent on the side of the aortic arch. Thus, in truncus arteriosus, the pulmonary artery is most frequently absent on the side of the aortic arch, in contrast to tetralogy of Fallot, in which the pulmonary artery is more frequently absent on the side opposite the aortic arch. Patients with truncus arteriosus are at risk of having pulmonary vascular obstructive disease develop at an early age, and this has provided the major impetus for early surgical correction [14]. however, pulmonary vascular disease tends to progress postoperatively more often in patients with single pulmonary artery than two artery [15].

This hazard of ventricular dysfunction may explain in part the observation that repair of truncus at 6 to 12 months of age is associated with a mortality twice that for repair between 6 weeks and 6 months of age [16]. The natural history of this condition is generally dismal. one series reported a survival of only 15% beyond the age of 1 year. Death is caused by heart failure, the complications of hypertensive pulmonary vascular disease and infective endocarditis. so early surgical intervention is advocated for these patients[17].

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Discussion :

Truncus arteriosus is a rare anomaly, defined as a single great artery that originates from the base of the heart and gives rise to the

pulmonary, systemic, and coronary circulation [18] .It is frequently associated with other cardiac anomalies including: VSD, aortic arch abnormalities, single ventricle, atrioventricular valve abnormalities, absent ductus arteriosus, and abnormal pulmonary venous return. Extracardiac anomalies occur in up 20% to 50% of the cases and include: situs inversus, asplenia, bony defects, cleft lip/palate, urinary tract abnormalities , absent gallbladder, hypoplastic lung, and neural tube defects. Patient in early infancy developed signs of heart failure , tachypnea , tachycardia , and failure to thrive(3. In this case with truncus arteriosus and increased pulmonary blood flow , may have to differentiated the condition from the other congenital cardiac conditions that cause early heart failure and are associated with either mild or no cyanosis. Such malformations include ventricular septal defect(Which is common cardiac lesions so the diagnosis didnt settled early) , patent ductus arteriosus, aorticopulmonary window, pulmonary atresia with ventricular septal defect and patent ductus arteriosus, or large collateral arteries, double-outlet right ventricle, univentricular heart, and total anomalous pulmonary venous connection.

- In any infant with mild cyanosis and signs and symptoms of congestive heart failure Truncus arteriosus should be suspected even it is a rare congenital cardiac malformation
- Operation should be done in first six month of age to avoid changes of pulmonary vascularity.

Recommendation:

1-It is crucial to diagnosis truncus arteriosus early in life to refer suspected case to tertiary cardiological centre.

2-Surgical operation shoud be doen as early as possible because delayed after six month of age increase mortality and morbidity.

3-Good preparation before surgery by using antifialure treatment and drug that lower pulmonary vascular resistance .

4-Follow up the patient after surgery by pediatric cardiologist looking for development of any complications.

In truncus arteriosus with decreased pulmonary flow, other conditions to be considered include pulmonary atresia, tricuspid atresia, tetralogy of Fallot, univentricular heart with pulmonary stenosis, and double-outlet right ventricle with pulmonary stenosis.

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ECG is non specific in diagnosis of trucus arteriosus(3).chest radiograph finding usually show cardiomegally ,plethoric lung, and right aortic arch is common.Echocardiograph reveal the origin of the pulmonary arteries,the truncal valve,VSD,and degree of overriding.

Color Doppler used for demonstration the regurgitation or stenosis of the trunca valve. cardiac catheterization with angiography used to define great vessels and coronary artery anatomy when there is suspicion of pulmonary vascular disease. sagittal and transverse MR image at the base of the heart can be used.

The prognosis is poor without treatment , Approximately 65% of untreated patient die before 6 months, and up to 90% die before one year of age.[8]

Early surgical repair(priorto6monthsofage) significantly improves outcome.[12,16]

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