CASE REPORTS

Tricuspid Atresia with Normal Axis on ECG Palliated With A Central Shunt; A Case Report

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Abstract

Background: *Tricuspid Atresia is the 3rd commonest cyanotic congenital Heart disease .It is characterized by lack of communication between the right atrium and right ventricle. The treatment often requires a palliative systemic to pulmonary shunt before definite surgery. The use of a central shunt via a median sternotomy is suggested here as an alternative to other traditional shunts via a thoracotomy.*

Method:*The management of a 3 month old boy who presented with dyspnoea, fever, cough and cyanosis is presented here as obtained from Clinical records.*

Result: *Following resuscitation, a central shunt (Ascending Aorta to main Pulmonary Artery) was constructed and the patient did well despite a turbulent post-operative period.*

Conclusion: *The management of tricuspid atresia likes other cyanotic heart disease is daunting but palliative treatment is possible in our environment and definitive treatment where possible affords a fairly satisfactory prognosis.*

Key words: *Tricuspid Atresia, Normal Axis, Central Shunt, Prognosis and Treatment.*

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Introduction

Tricuspid Atresia (T.A.) constitutes about 3% of congenital Heart disease[1].It is the 3rd commonest cyanotic heart disease[2, 9]. The aetiology of T.A. is unknown[1]. Symptoms of T. A. manifest early in life. Nearly one half low pulmonary blood flow (oligaemia) present early with cyanosis while those with high (plethora) flow may present later with congestive heart failure[3].

Treatment begins usually with patient stabilization including preventing ductal closure with prostaglandin E_1. This is usually followed with a systemic to pulmonary artery-shunt (e.g. modified Blalock-Taussig, central shunt etc) if patient presents by 3 months or a systemic venous to pulmonary artery shunt if patient presents later. The use of 2-D Echocardiography is often sufficient [2,4]for diagnosis while cardiac catheterization may be required before definite treatment where the anatomy or an aspect of the patient cardiac physiology needs to be clarified[4]. Without treatment only 10% are alive by 10yrs but this can be increased to about 81% with early and appropriate treatment[1,3].

We present the case of a 3month old boy with tricuspid atresia palliated with a central shunt via a median sternotomy rather than the more conventional Modified Blalock-Taussig shunt via a posterolateral thoracotomy with a remarkable outcome.

Case Report

J.A presented at two months with one month history of difficulty with breathing three days history of cough and fever. He is a product of full term supervised pregnancy and was delivered spontaneously per vagina. The mother ingested a local concoction at three months of pregnancy consequent upon which she had diarrhoea (8 episodes over 2 days) and intestinal colic .He was well until a month after delivery when the mother noticed above symptoms in addition to baby turning blue/dark on exertion or while crying. No other congenital anomaly was noted. He was earlier seen at two peripheral hospitals before referral to the University of Nigeria Teaching Hospital Enugu where he was managed by the paediatricians for 2 weeks before referral to the Cardiothoracic unit as a result of worsening symptoms. While with the paediatricians he had repeated episodes of apnoeic spells and they made the diagnosis of T.A from 2-D Echocardiography. Physical Examination revealed a dyspnoeic and cyanotic baby boy on oxygen (o_2) by nasal catheter with SPO_2 (O_2 saturation) of 70% on O_2 and between 40-50% on room air. He had extensive soft tissue necrosis over the extensor surface of the right wrist(2% body surface area) dressed with gentian violet solution, a consequence of hypertonic dextrose infusion. His Heart

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rate was 160/minute, Heart sound I and II and a systolic murmur 3/6 maximal at the upper left sternal border. Result of investigation revealed a packed cell volume of 58%.Other biochemical and hematological indices were unremarkable. Chest X-ray revealed pulmonary oligaemia 2-D echocardiography revealed atretic tricuspid valve , hypoplastic right ventricle ASD and PDA, while ECG revealed bi-atrial enlargement with normal axis(see figure 2). Patient was duely resuscitated and prepared for surgery. Via a median sternotomy the mediastinum was opened and the Ascending.
Aorta was connected to the Pulmonary trunk using a 5mm Gor-Tex graft

(Polytetrafluoroethylene);CENTRAL SHUNT .Post-operatively His Saturation Improved To >90% on O_2.He developed superficial wound dehiscence noticed on the 16th postoperative day which was aggressively managed by wound debridement ,daily wound irrigation with antibiotic solution and wound dressing. He was discharged to the surgical out patient unit on the 65th post-operative day with SPO_2 of 89% on room air and a weight of 6Kg (85% of expected).

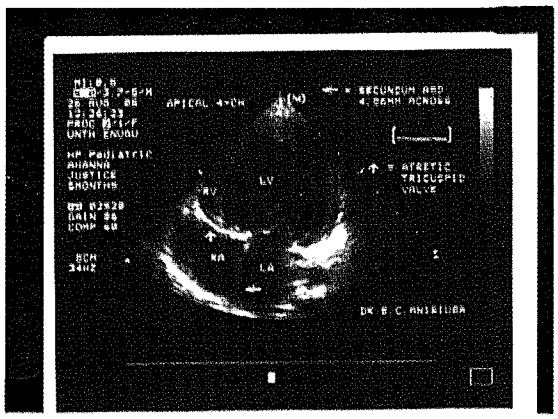

Figure1. 2-D Echo (Apical 4-Chamber view) Showing Atretic Tricuspid Valve (?), Hypoplastic Right Ventricle (RV) and Secundum ASD (?)

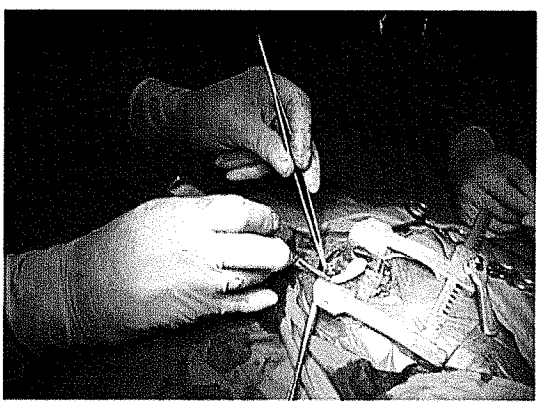

Figure 2.Intra-Operatively; Surgeon Pointing To Central Shunt (Whitish) between Blades of Sternal Retractor

Discussion

T.A is a cyanotic congenital heart disease characterized by lack of communication between the right atrium and the right ventricle. Associated anomalies include atrial septal defect, patent ductus arteriosus, and hypoplastic right ventricle [3,4]. Occasionally in about 30% of cases a more complex condition occurs in which there may be transposition of the great arteries (Types II and III) and varying degrees of mitral valve and left ventricular enlargement. Our patient had the typical T.A (see figure 1)

It is the 3rd commonest cyanotic heart leision. The aetiology of T.A is unknown. A multifactorial inheritance hypothesis is offered to explain the aetiology in which genetic and environmental factors interact during a critical period of cardiac morphogenesis however no specific factors are clearly identified for T.A[1]. Could the concoction the mother ingested at 3-month of gestation have any causal relationship with this pathology?

The pathophysiology is dependent on the degree of obstruction and pulmonary blood flow. Cyanosis is present at birth in 71% of patients or appear later .Late presentation appears when the patent ductus arteriosus begins to close. Presentation is with dyspnoea, fatigue, cyanosis, apnoeic spells, retarded growth and development and clubbing in older children. Except for the latter these features of failure to thrive were present in A.J. Other findings include prominent jugular veins strong left ventricular impulse ventricular septal defect. Chest X-ray typically shows oligaemic lung field but may be plethoric if associated with transposition of the great arteries. ECG shows bi-atrial enlargement, left ventricular enlargement and left axis deviation. This patient however had a normal axis which is unsual. The usual presentation is an abnormal superior vector (left axis deviation 0^0 to -90^0 in the frontal plane).Normal axis (0^0 to $+90^0$) or right axis ($+90^0$ to $+/-180^0$) is present in a minority of patients mainly in T.A. types II or III. A number of mechanisms have been postulated to explain the abnormal superior vector including destructive lesions on the left anterior bundle, fibrosis of the left bundle branch, a long right bundle branch along with an early origin of left bundle branch, a small right ventricle and a large left ventricle. Data from recent ventricular activation studies suggest that the superior vector is likely due to the interaction of several factors[1]. While the literatures are mute on the pathogenesis of the normal or right axis pattern, it can only be surmised that the absence of the factors which interact to produce the abnormal superior vector may

allow for a normal or right axis deviation. 2-D echocardiography is usually sufficient for diagnosis while cardiac catheterization ± angiography may be required before the definitive procedure to define more complex anatomies[1,4]. Patients less than 3 months are usually given systemic to pulmonary artery shunts (e.g. modified Blalock-Taussig) due to the persistence of the high fetal pulmonary pressure which usually drops after 3 months allowing for the construction of a systemic venous to pulmonary artery shunt(Glenn or bi-directional Glenn). The definitive surgery is indicated for severe/progressive cyanosis and a high/rising hematocrit (>50%). Traditionally palliative procedures are done through a thoracotomy because this leaves a virgin sternum for the definitive surgery via a median sternotomy[4]. We chose the latter approach because of its advantages and the current trend. The advantages include excellent palliation of cyanosis, no distortion of branch or peripheral pulmonary arteries, high shunt patency rate, equal blood flow to the lungs thus avoiding asymmetric growth of both lungs and it is easier to take down during the definitive surgery[5,6]. One major disadvantage is difficulty with regulating blood flow which may eventually lead to early pulmonary hypertension if flow is excessive. Also Odim et al have shown that shunts done through a sternotomy route are technically less challenging and it is associated with fewer shunt failures than the classic thoracotomy approach[6,7]. The definitive treatment is by the various modification of the Fontan procedure in which systemic venous blood is channeled to the pulmonary artery with ASD closure. This done between 2-5years[1,8]. The 5, 10 and 15 years Survival are approximately 86, 81 and 73% respectively[8].

Conclusion

T.A with a normal axis is uncommon and this is the first reported case of one palliated with a central shunt in this sub-region. The remarkable outcome lends credence to recent claims in western literatures on the versatility of the central shunt and the safety and ease of the sternotomy approach[1,5]. Therefore local practitioners should be familiar with the presentation and management of T.A and other cyanotic heart conditions and refer early to tertiary centres equipped to handle such cases.

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