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Good Prognosis of ALCAPA (Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery Syndrome) with early Diagnosis and Surgical Treatment

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Abstract- Introduction: ALCAPA Anomalous origin of the left main coronary artery (LCA) from the pulmonary artery (PA) is a rare malformation that presents with myocardial ischemia or infarction and/or cardiac failure in infants (1). Surgical correction to re-establish a two-coronary artery perfusion system is the treatment of choice (2).

Objective: To present a case of ALCAPA with early diagnosis and treatment, and a very good prognosis.

Case Relate: 6 months old girl admitted with symptoms of heart failure, Echo, ECG and Cath showed signs of ALCAPA with sever dilated cardiomyopathy. The baby sent for surgical repair, and direct implantation of the LCA into the ascending aorta was done successfully. The baby followed in our cardiology clinic monthly. After 6 month of surgical repair, the baby was good without any complain, all symptoms resolved, the Echo showed acceptable contractility and function, the dilated cardiomyopathy improved dramatically.

Conclusion: In infants with DCM due to coronary artery anomalies, early diagnosis and surgical treatment with optimal timing provide an excellent prognosis.

Keywords: ALCAPA, dilated cardiomyopathy, re-implantation of the coronary artery.

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I. INTRODUCTION

ALCAPA Anomalous origin of the left main coronary artery (LCA) from the pulmonary artery (PA) is a rare malformation also known as Bland-White-Garland syndrome that presents with myocardial ischemia or infarction and/or cardiac failure in infants(1). It is associated with a mortality rate of 90% within the first year of life. Surgical correction to re-establish a two-coronary artery perfusion system is the treatment of choice. So we will present a case of ALCAPA which corrected successfully by surgery (2).

II. CASE REPORT

6 months old girl admitted to the hospital with cough, breathing difficulty and sweating especially during crying or feeding from about 3 months, these symptoms were mild in the beginning but then became worse gradually, and associated with poor gaining weight.

She was full term baby, normal delivery without any complications, birth weight: 3.8kg.

On physical examination: tachypnea RR = 45/min, Failure to thrive (WT=4.9kg <3%, HT=55cm <3%), Soft systolic murmur III/VI at the apex, chest: Harsh crackles bilateral Liver palpable about 4 cm below costal margin, other systemic examination was normal Labs: WBC: 11000 (N 35%, L 62%), Hgb 11.2 g/dl, CRP negative Electrolytes, liver function, renal function and Vit. D were normal.

Capillary Blood gases: PH = 7.51, PCO₂ = 32, PO₂ = 86, Hco₃ = 26, So₂ = 97% Chest XR: Cardiomegaly, increased pulmonary vascularity bilateral (Figure 1).



Fig. 1: Chest XR: Cardiomegaly, Increased Pulmonary Vascularity Bilateral

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ECG: Signs of Anterolateral Myocardial Infarction (ST Depression in V5, V6) (Figure 2, 3).

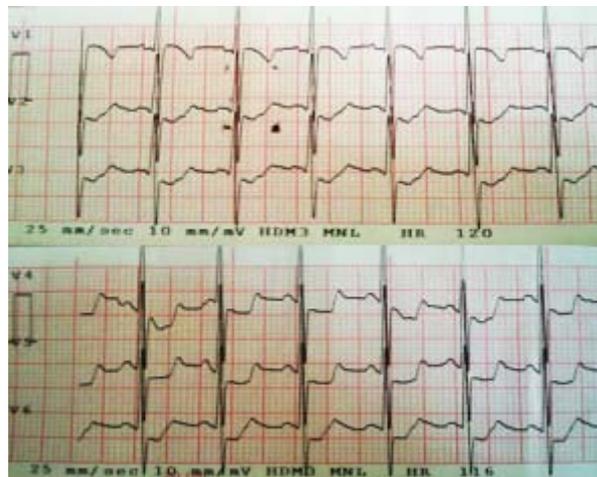
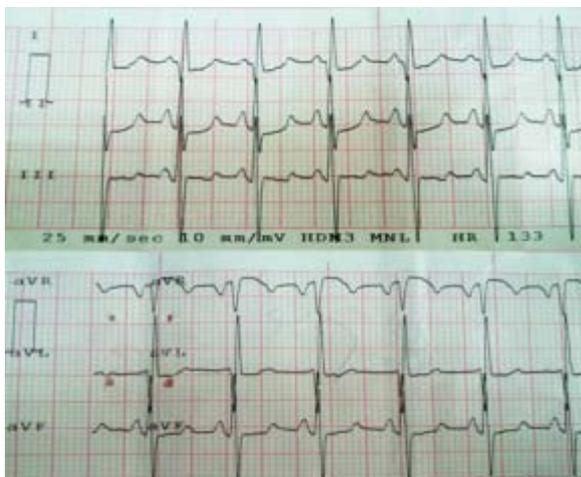


Fig. 2, 3: ECG: ST Depression in V5, V6.

ECHO cardiogram: poor function, sever dilated left heart, LVEDd = 6cm, EF = 14%, Fs = 7%, moderate to severe mitral regurgitation , left coronary artery cannot be seen from the Aorta , but seen as a diastolic flow in the main pulmonary artery Conclusion: sever dilated cardiomyopathy, suspected ALCAPA (Figure 4, 5).

So we started heart failure medications and sent her to the cathlab to complete the diagnosis.

Cath showed only RCA came from Aorta (AO) (Figure 6), LCA from Pulmonary artery.

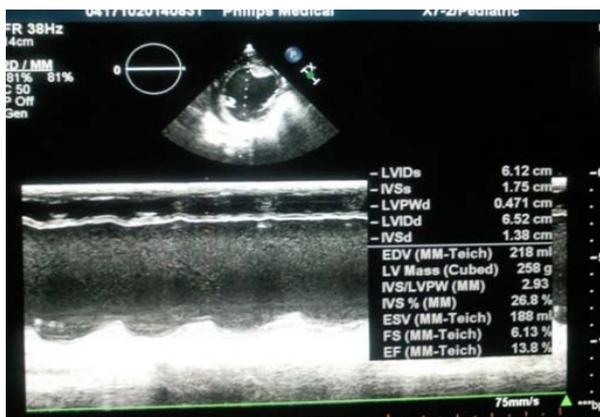


Fig. 4: Echo: M Mode in the LV Showed the Sever Dilatation and Poor Function



Fig. 6: Cath Showed the Aorta, only the RCA (Right Coronary Artery) Came from it

The baby sent for surgical repair, and direct implantation of the LCA into the ascending aorta was done successfully.

The Echo after surgery showed LCA came from Aorta as in the normal heart (Figure 7).

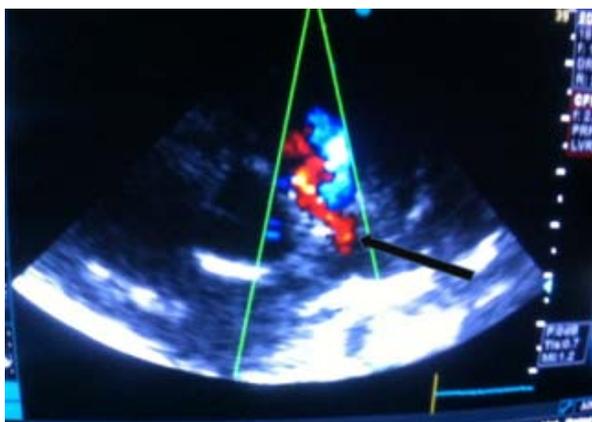


Fig. 5: Echo: Short Axis View Showed the Left Coronary Artery Origin from the Pulmonary Artery



Fig. 7: Echo Post-Surgery: Left Coronary Artery Origin from Aorta

We continued the heart failure medications after surgery for 3 months, the baby followed in our cardiology clinic monthly.

After 6 month of surgical repair, the baby was good without any complain, all symptoms resolved, WT = 8.9 kg.

The Echo showed acceptable contractility and function, the dilated cardiomyopathy improved, EF became about normal EF = 52% (Figure 8).

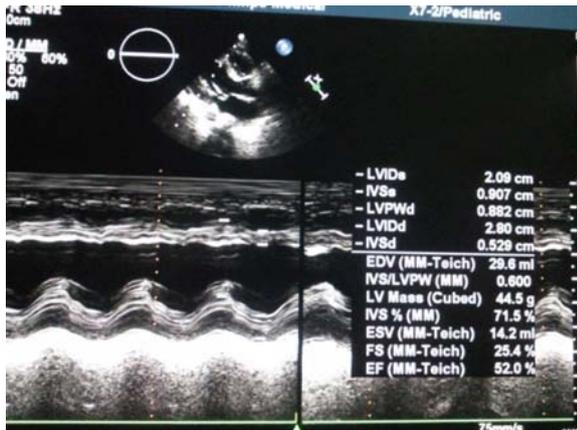


Fig. 8: Echo: 6 Months Post-Surgery, Good Contractility

III. DISCUSSION

ALCAPA is a rare but serious congenital anomaly. Presently, the prognosis for patients with ALCAPA is dramatically improved as a result of both early diagnosis and treatment, The presenting features are paroxysms of irritability, which correlate with signs of heart failure so dilated cardiomyopathy is an important differential diagnosis and may also arise as a result of ALCAPA (3) as in our case which presented with dilated cardiomyopathy symptoms (4).

During infancy, ALCAPA syndrome can be fatal and patients present with myocardial infarction, left ventricular dysfunction, mitral regurgitation, or silent myocardial ischemia, which can lead to sudden cardiac death (4). While adult patients can be completely asymptomatic, they may present with angina, dyspnea, syncope, myocardial infarction, arrhythmia or sudden cardiac death (5). The electrocardiogram of a baby with ALCAPA usually shows typical signs of an anterolateral myocardial infarction, manifested by abnormal Q waves in leads I, AVL, V5, and V6, as well as by transient ST changes in these leads (we saw these changes in our case). The ECG may be also entirely within normal limits (6).

In patients with ALCAPA syndrome, even if the patient is asymptomatic, or symptomatic, surgical treatment is suggested. In our case, the patient was operated as soon as the diagnosis was reached. The objective of surgical treatment is to restore a normal coronary circulation and improve myocardial perfusion. A few surgical procedures are suggested based on the

localization of the abnormal coronary artery ostium. In our case the LCA was directly re-implanted to the Ao. After surgery regular follow up is mandatory especially during the recovery period because of arrhythmia risk and sudden death. Early treatment may prevent irreversible myocardial damage with its subsequent complications (7), and we saw that in our case which dramatically improved after surgical repair.

IV. CONCLUSION

In infants with DCM, coronary artery anomalies should be considered a priority in establishing a diagnosis. Coronary re-implantation is the technique of choice for surgical correction. Early diagnosis and surgical treatment with optimal timing provide an excellent prognosis.

Abbreviations

ALCAPA : Anomalous origin of the Left Coronary Artery from the Aulmonary Artery.

Ao : Aorta. DCM : Dilated Cardiomyopathy.

PA : Pulmonary Artery. ECG : Electrocardiography.

EF : Ejection Fraction. LCA : Left Coronary Artery.

RCA : Right Coronary Artery.

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