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Case Report

Common Atrium Concurrent with Atrioventricular Septal Defect: A Case Report

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Abstract: Common atrium accounts for 0.5% to 1% of congenital heart diseases. It is characterized by the atria septum's complete absence and atrioventricular canal defect. It may occur as an isolated malformation or associated with other extracardiac anomalies. Untreated cases are at risk of developing pulmonary hypertension. We present a case of an 18-month-old female baby with a recurrent chest infection, central cyanosis, and echocardiographic features of the common atrium concurrent with atrioventricular canal defect and features of pulmonary hypertension.

Keywords: Common atrium, atrioventricular septal defect [AVCD], Congenital heart diseases [CHD], Echocardiography, Dual-Source Computed Tomography [DSCT].

BACKGROUND

Common atrium is a condition characterized by the complete absence of interatrial septum and malformation of the atrioventricular [AV] valves with or without interventricular communication.it is not synonymous with a single atrium which is characterized by i) complete absence of interatrial septum, ii) absence of malformations of AV valves, and iii) absence of interventricular communication as recommended by (Levy, Salomon, and Vidne 1974; Franklin *et al*., 2017).

Common atrium is a complex and rare congenital heart malformation accounting for 0.5% to 1% of congenital heart diseases (Campbell 1973). Usually but not always, may be associated with or without extracardiac vascular malformations (Garg and Jain 2021; Zhang *et al*., 2018).

It can occur as an isolated entity or syndromic with Ellis-van Creveld syndrome, heterotaxy syndrome with asplenia, and trisomy 21 but is rare in non-syndromic clients (Ferdman, Brady, and Rosenzweig 2011; Kim *et al*., 2016).

Patients with common atrium are at increased risk of developing pulmonary hypertension. However, some patients develop pulmonary hypertension earlier than others, the reason for this variability is unknown, and determining which patient will develop pulmonary hypertension earlier than others is still challenging (Ferdman, Brady, and Rosenzweig 2011).

Echocardiography is the first-line imaging modality to diagnose this condition (Zhang *et al*., 2018) and noninvasively determine whether the client has pulmonary hypertension (Agarwal *et al*., 2021). Here, we report a case of a common atrium concurrent with an atrioventricular canal defect [AVCD] and echocardiographic features suggestive of pulmonary hypertension.

CASE PRESENTATION

An 18-month-old female bay was brought for a chest x-ray to evaluate the causes of recurrent cough. She has been experiencing recurrent cough since the age of 9 months, recently it has progressively worsened in the past month despite the use of several antibiotics

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including ceftriaxone and over-the-counter, amoxicillin, and cough syrups. Occasional swelling of the eyelids in the morning. She was born at term by a healthy mother without antenatal or perinatal complications with a birth weight of 3.4kg, at the time of presentation to the hospital her body weight was 8.2kg which dropped from 10.6kg recorded 3 months before this visit, her early milestones were normal sitting at 6monhs and crawling at 8-9months but delayed to stand up without support which occurred at 1year and 3months. Her vaccination history was up to date according to the Tanzania immunization schedule.

On examination, she was conscious with central cyanosis, mild finger clubbing, jugular venous distention and pulsations, oxygenation saturation of 73% to 76% without oxygen, and up to 89% to 90% with oxygen supplementation, irritability, and her weight was within the green zone of the growth chart. Auscultation demonstrated bilateral basal crepitation and a holosystolic murmur at the apex. There was no lower limb swelling.

Chest x-ray: showed cardiomegaly with a cardiothoracic ratio of 0.85, prominent artery, and pulmonary edema (Figure 1).

Echocardiography revealed a visceral situs with levocardia, a large common atrium [173cc]. With the right side, demonstrating right atrial morphology[inferior vena cava drains into the right side of the atrium] and the left side demonstrating left atrial morphology [presence of left atria appendage and draining pulmonary veins], a single atrioventricular valve [AVV] with two valvular orifices[mitral and tricuspid orifices] and severe regurgitation(covering more than 50% of the common atria area) (Figure 2) with a pressure gradient of PGmax:78.29mmHg predominantly from the tricuspid orifice, Moderate right ventricular enlargement RV=LV [Figure 3]. Normal aortopulmonary Orientation [crossover], Aorta diameter [13.5mm], dilated main pulmonary artery diameter [MPA]- 20.2mm, right[13.4mm] and left[12.6mm] pulmonary arteries [MPA/AO=1.5] impressive of pulmonary over circulation/pulmonary hypertension [figure 4A, B]. Global good ventricular contractility with left ventricular ejection fraction [EF] of 90% (by Teichholz method) Figure 4C. Rounded, dilated IVC-13.2mm, and hepatic veins [right hepatic vein (RHV-11.3mm), left hepatic vein (LHV-8.8mm), Middle hepatic vein (MHV-8.8mm)] (Figure 5A, B).

Treatment included; Furosemide 10mg once daily, captopril 6.25mg once daily, and spironolactone 6.25mg once daily and was referred to a specialized cardiac Centre [Jakaya Kikwete Cardiac Institute], whereas of the last follow-up she was provided with the same medications but slight change in dosage and frequency of captopril 3.125mg thrice daily, furosemide 10mg twice daily and spironolactone 6.25mg twice daily. The mother reported an improvement in symptoms and was scheduled to return to the specialized cardiac center after three months.

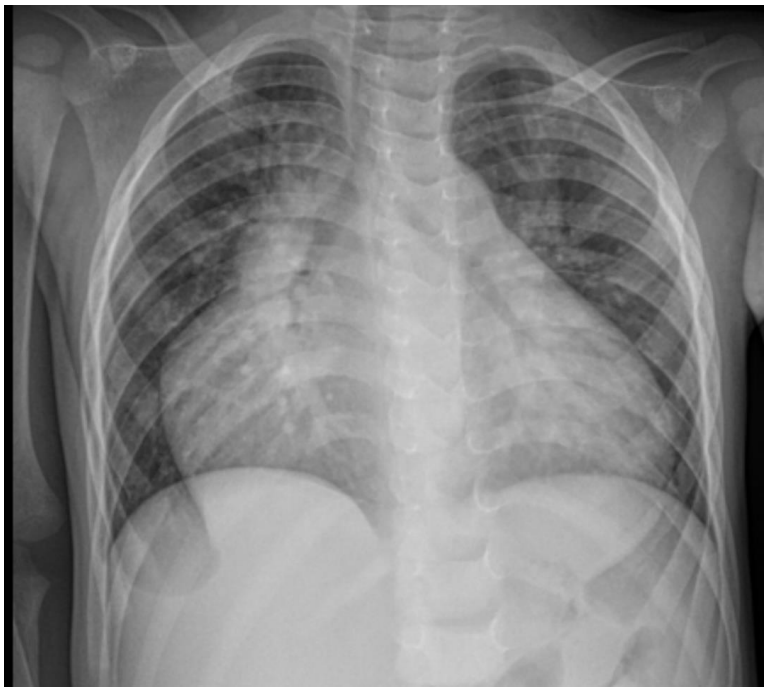

Figure 1: PA chest radiography demonstrating, cardiomegaly (CTR-0.85) and pulmonary edema

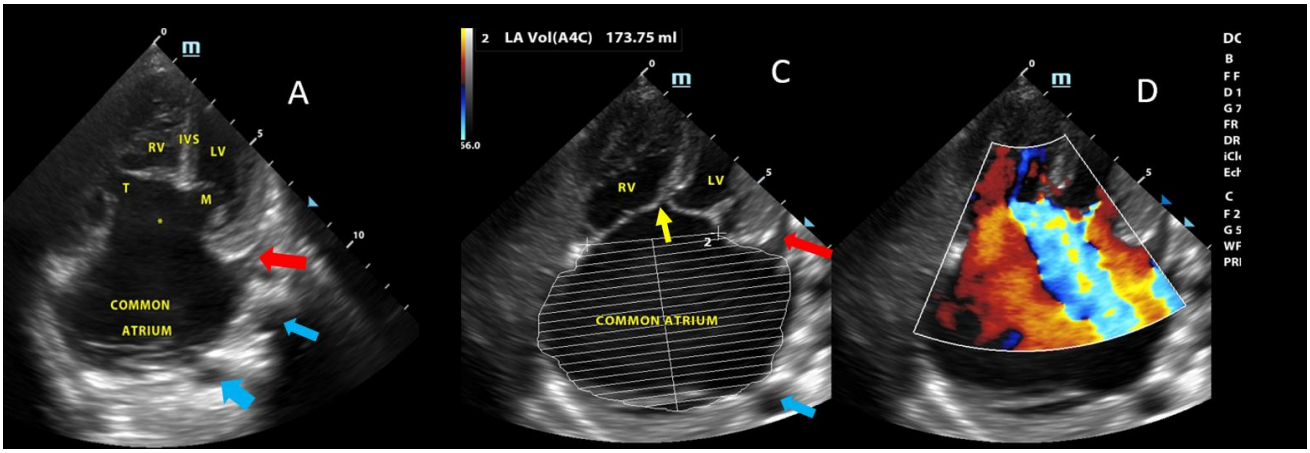

Figure 2: 2D(A&B) and color flow mapping(C), Apical 3 chamber views (four-chamber views for the normal heart), A: showing common atrium, common atrioventricular canal defect [AVCD] *, two valve orifice [m-mitral orifice and T-tricuspid orifice], blue arrows-pulmonary veins, red arrow-left atrial appendage, B: common atrium volume [173cc], relatively same sized right ventricle [RV] and left ventricle [LV], same plane of atrioventricular attachment to the septum [yellow arrow], C: severe eccentric atrioventricular valve regurgitation predominantly towards the left side

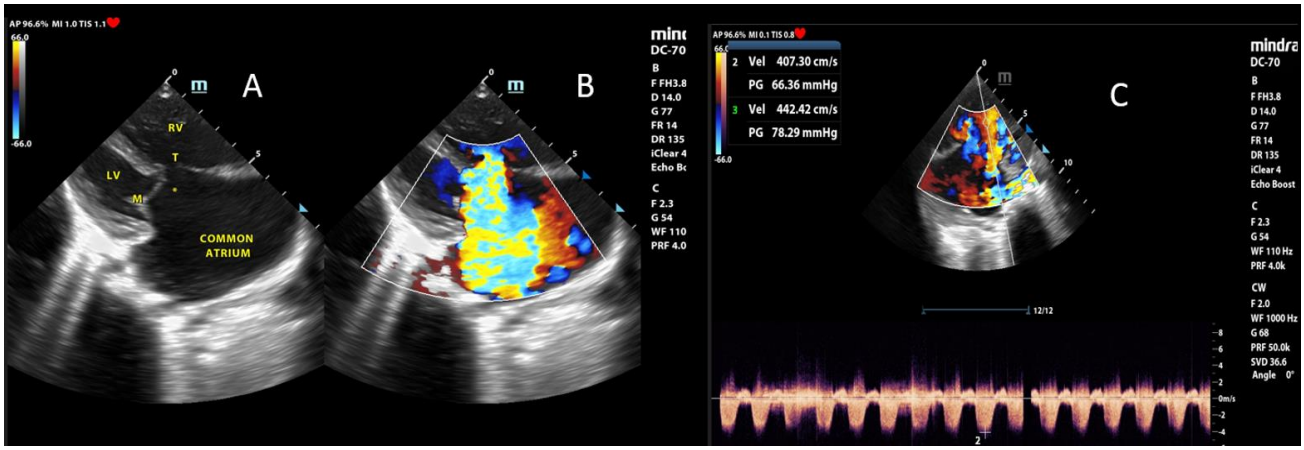

Figure 3: Parasternal Long Axis views, 2D(A) showing common atrium, common atrioventricular canal defect [AVCD] *, two valve orifice [m-mitral orifice and T-tricuspid orifice], flattened septum and slightly large RV than LV. (B) color flow mapping showing severe eccentric atrioventricular valve regurgitation covering approximately more than 50% of the common atrium and directed towards the posterior wall of the common atrium and color Doppler (C) showing tricuspid regurgitation maximum velocity 4.42m/sec and pressure gradient PG 78.29mmHg

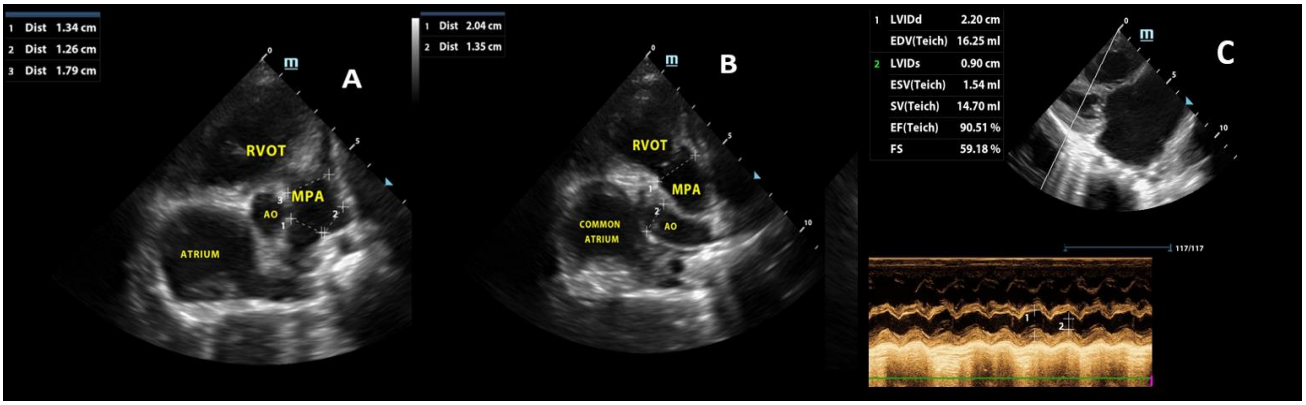

Figure 4: 2D Echocardiogram, [A&B]short axis aortal pulmonary view: Showing main pulmonary [MPA] and its relation with the Aorta [the normal crossover]], the main pulmonary artery diameter [17.9-20.4mm] is larger than the Aorta[13.5mm, the pulmonary artery to aortic ratio is 1.5, the right[RT-13.4mm] and left[LT-12.6mm] pulmonary artery are also dilated ,C] Mmode parasternal long axis view showing ventricular interdependence and ejection fraction of 90%

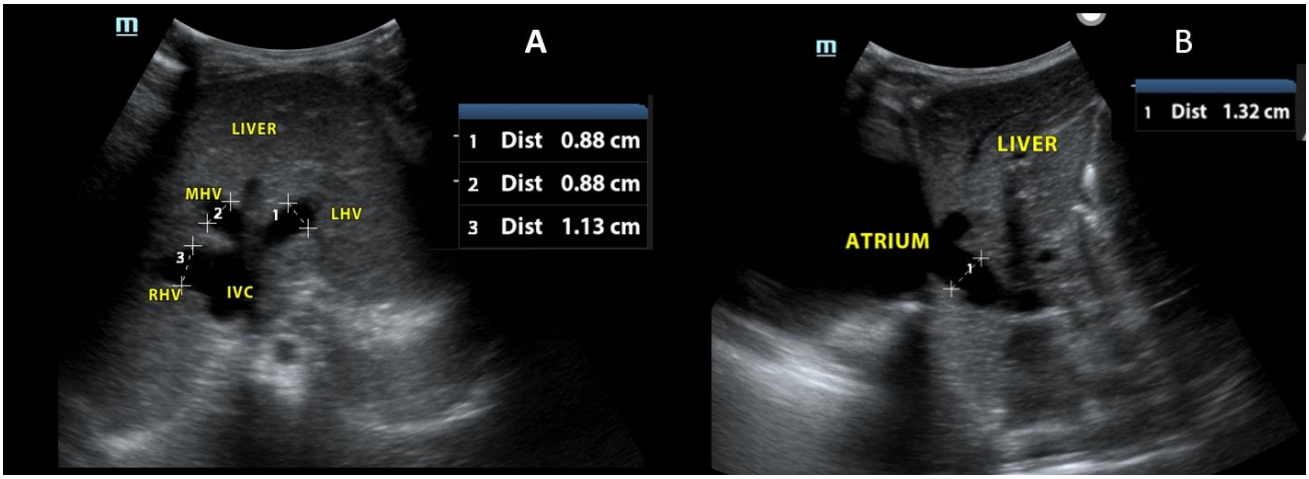

Figure 5: 2D sonogram transverse view of the liver through the confluence of the hepatic veins to the inferior vena cava(A), The hepatic veins are dilated, LHV-8.8mm, MHV-8.8mm, RHV-11.3mm. sagittal view of the liver demonstrating the IVC and its diameter-13.2mm], LVH-left hepatic veins, RHV-right hepatic vein, MHV-middle hepatic vein

DISCUSSION

Common atrium is a rare congenital heart disease; most cases in the literature are case reports or case series (Agarwal *et al*., 2021; Rastelli *et al*., 1968; Zhang *et al*., 2018). The first case was reported in 1907 by (Young 1907). Since then, there has been confusion and inconsistency in the nomenclatures of this condition in human and veterinary literature, as some authors described it as a variant of partial atrioventricular canal defect [partial AVCD] or common atrium with AVCD. However, this does not precisely fit the description since in an atrioventricular canal defect there is at least a component of atrioseptal structure on top of the interatrial communication (Agarwal *et al*., 2021). This implies that our case fits well with the term common atrium concurrent with AVCD.

It is clear from the literature that Correct and consistent nomenclature for congenital heart disease [CHD] is necessary for precise and accurate communication among colleagues and very important for surgical planning and prediction of outcomes of surgical repair. However; due to the unusual spectrum of presentation and associated anomalies in congenital heart diseases, it's important not to rely on the names of the particular diseases rather than on the precise description of the findings seen during imaging examinations to aid in case management (Agarwal *et al*., 2021). Our case has a common atrium and atrioventricular canal defect demonstrating the unusual appearance of congenital heart disease. In this case, the description of echocardiographic findings is important for medical and surgical planning rather than relying on a single term to name the case.

Clinically patients present with fatigue, palpitations, exertional dyspnea, cyanosis, recurrent upper respiratory infections, and delayed developmental milestones, especially in their childhood. Due to the earlier presentation of symptoms, most diagnoses are made during childhood (Azaje *et al*., 2023; Kim *et al*., 2016). However, in resource-limited settings, early diagnosis is partly dependent on a high index of suspicion by the attending clinician prompting him or her to order echocardiography. In our client, the decision to perform echocardiography came after the chest x-ray showed cardiomegaly otherwise she has been frequently treated as a case of recurrent pneumonia.

Of note; there are reported cases of clients who survive to adulthood before the diagnosis is made, The oldest patient with a common atrium reported in the literature is a 48-year-old male (Kim *et al*., 2016). The question is why some grow without or with minimal symptoms to adulthood and others do not. The possible explanation could be the existence of variability in hemodynamics among clients with some clients having favorable hemodynamics and others unfavorable ones or in part due to variability in the coexistence of extracardiac vascular abnormalities that confound the hemodynamics and therefore the severity of symptoms.

(Azaje *et al*., 2023; Rastelli *et al*., 1968) suggest that despite the mixing of arterial and venous blood in the common atrium which is almost similar to that of a large atrioseptal defect the most important factor that determines symptoms is the status of the AV valve and the predominance of direction when blood exits the atrioventricular canal resulting into some client to have more oxygenated systemic circulation than pulmonary circulation and hence those with more oxygenated systemic circulation tend to experience mild symptoms and delay in the diagnosis .our client developed symptoms earlier than most of the reported cases due to low oxygenation of the systemic circulation which is partly contributed by severe AV valve regurgitation (Figure 2D). Furthermore; (Rastelli *et al*., 1968) suggests that regurgitation increases the mixing of blood in the common atrium thereby decreasing oxygenation of the systemic circulation. In addition, regurgitation causes severe pressure and volume overload resulting in

enlargement of the common atrium. In our case, the common atrium measured 173cc which is relatively higher than the combined volume of the adult right and left atrium and probably the largest common atrium reported in the literature. The regurgitation has further increased the backward pressure transmitted in the inferior vena cave and hepatic veins causing them to dilate (Figure 5A, B) and superior vena cava, which explains the pulsations and distention of the jugular veins. Since the atrium has tremendously increased in volume, this might probably affect the decision to perform surgery if planned in the future.

Echocardiography is the first-line imaging modality to diagnose this condition and offers an advantage in determining cardiac functions and pulmonary hypertension (Kim *et al.*, 2016; Zhang *et al*., 2018). It is well documented that the common atrium is frequently accompanied by congenital abnormalities of systemic and pulmonary drainage [extra cardiac vascular malformations] (Zhang *et al*., 2018). In our case, extracardiac anomalies were not evaluated due to the limited examination window of echocardiography.

Currently, low doses of dual-source CT scans have proven to be the imaging modality of choice in the diagnosis of extracardiac malformations accompanying common atrium, In their study (Zhang *et al*., 2018) demonstrated that the sensitivity of dual-source computed tomography [DSCT] is higher than transthoracic echocardiography [TTE] in the detection of extracardiac vascular malformations accompanying common atrium [i.e. 92.31% vs 76.92% respectively] due to high resolution and more post-processing options with CT than Echocardiography. The authors recommend computed tomography [CT] as the best alternative for evaluating congenital heart diseases.

In our case, we cannot state confidently that there were no associated extracardiac abnormalities, as a CT scan was not performed due to its limited availability in our setting and caregivers' financial constraints. However, echocardiography has shown the presence of pulmonary hypertension as demonstrated by an increased pulmonary artery-to-aortic ratio [Figure 4A, B]. It is documented that; a pulmonary-to-aortic ratio of >1:1 is a strong predictor for the presence and severity of pulmonary hypertension (Schneider *et al*., 2020). In our case, the ratio was 1.5:1 impressive of the existence of pulmonary hypertension.

Surgery is the treatment of choice for Treatment of the common atrium and it is preferably done earlier since the overall prognosis is poor in early symptomatic cases due to the development of pulmonary hypertension and cardiac dysrhythmias (Kim *et al*., 2016; Ferdman, Brady, and Rosenzweig 2011)**.** Our client was managed medically and referred to Specialized Cardiac Centre [Jakaya Kikwete Cardiac Institute], follow up will be made after three months.

CONCLUSION

Common atrium is rare, it is not synonymous with single atrium, we have described a case of common atrium concurrent with an atrioventricular canal defect who presented with a recurrent chest infection. The early presentation of symptoms is due to severe valvular regurgitation and early development of pulmonary hypertension. Early diagnosis is important for early treatment planning. Once diagnosed a further search for other congenital anomalies should be made using Dual-Source Computed Tomography [DSCT], as there is an increased association with other cardiac and extra-cardiac vascular malformations.

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Conflict of Interest: There are no conflicts of interest

Informed Consent: Consent was obtained from the client's parents

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