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Congestive heart failure in children with atrial septal defect: a case report

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The aim of this study is to present a clinical case referred to a 12-year-old boy with history of atrial septal defect (ASD) and HF shows a response to treatment with improved symptoms and quality of life. The patient's laboratory studies shown increasing of neutrophil lymphocyte ratio. Chest X-ray showed a cardiomegaly. The patient had no peripheral oedema. The echocardiography identified there were a 1.88-cm ostium secundum atrial septal defect with right atrium, and right ventricle dilatation. This case highlights the associated between congenital heart disease and heart failure in paediatrics. It also illustrates exercise intolerance could be a presentation of undiagnosed atrial septal defect with heart failure. The conclusion is the patient had developed symptomatic HF with LVEF ≥ 50 % as well as RA and RV dilatation. Most likely this patient developed HF due to the delayed diagnosis of ASD.

Keywords: heart failure, child, atrial septal defect, congenital heart defects.

Conflict of interests: none declared

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Introduction

Atrial septal defect (ASD) is condition of the wall (septum) that divides atrium are opened [1]. Those condition are one of the most common congenital heart defects (CHD) occurred in about 25 % children and estimated incidence of 56 per 100,000 live births [2–4]. Small defects on atrial septal usually can be closed spontaneously in childhood, whereas large de-

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fects can cause complications such as heart failure, dysrhythmias, and pulmonary hypertension. Doing a surgical intervention is necessary to prevent those complications in children [1].

Congestive heart failure (CHF) sometimes known as heart failure (HF). HF is defined as the clinical syndrome associated by the cardiac structure or function abnormalities. Paediatrics HF is rare than HF in adult. Although relatively rare, CHD and cardiomyopathies are the most common aetiologies of HF in children [5]. Epidemiology of HF in children isn't well characterized, but in United States (US) the incidence of HF occurred in children is 0.97 to 7.4 per 11,000–14,000. [6] The aetiologies of HF in CHD can be divided into 2 conditions of pathophysiology, there are: (1) over circulation failure; and (2) pump failure [7].

HF intervention in the setting of CHD is challenged. The aim of this research is to present a clinical case referred to a 12-year-old boy with history of ASD and HF shows the response to treatment with improved symptoms and quality of life.

Case Report

A 12-year-old-boy was admitted on the emergency department (ED) complaining of the sudden onset of dyspnoea that appeared after activity and lasted for several hours. Dyspnoea appeared suddenly and lasts continuously before entering the hospital. His complaint didn't improve at rest. There were minimal intercostal retractions, but there are no abnormal breath sounds were described on admission. On examination, there was a soft mid-systolic murmur at his upper left sternal. The patient was an elementary school's student and reported his activity more fully than before. There were no chest tightness or palpitation, and respiratory symptoms. Previously, the patient presented with the sudden onset of shortness of breath after doing sport, but it was improved at rest. In addition, he also complained of considerable orthopnea while lying down, which was relieved by sleeping prone, and frequent shortness of breath and colds. There was no family history of cardiovascular events or deaths in young people.

He had a heart rate of 103 bpm, blood pressure of 93/57 mmHg, a respiratory rate of 23 breaths per minute, an oxygen saturation of 100 % on 2 lpm of oxygen. Examination revealed clean lungs and a faint mid-systolic murmur at his upper left sternal. There weren't both of ascites, and oedema on his extremities. The

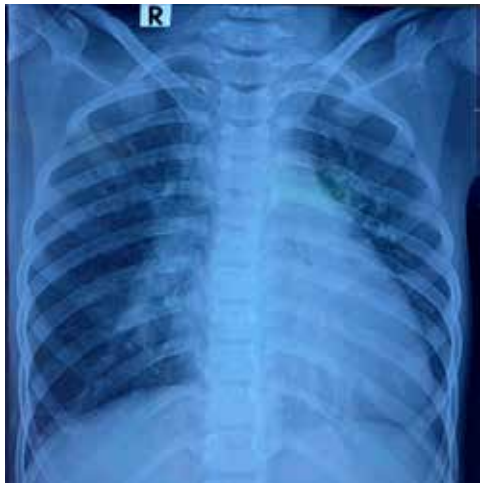

Fig. 1. Chest X-Ray P/A view showing cardiomegaly with Cardiothoracic ratio (CTR) 0.59

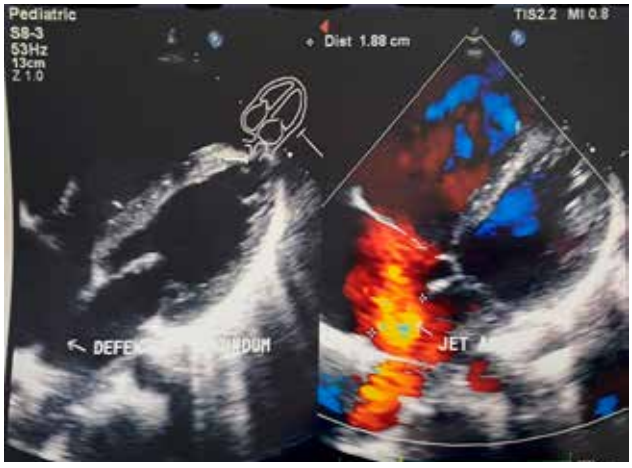

Fig. 2. Echocardiography showing the ostium secundum ASD (Diameter 1.88 cm)

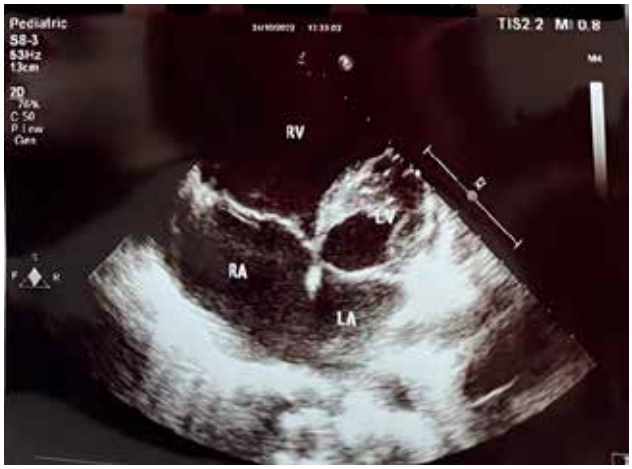

Fig. 3. Echocardiography showing RA-RV dilatation

result of his laboratory studies shown increasing of neutrophil lymphocyte ratio (NLR). Chest X-ray (CXR) on anteroposterior view (Figure 1). Based on the CXR results, the patient did an echocardiography examination, and the result shown in (Figure 2, and 3). CXR,

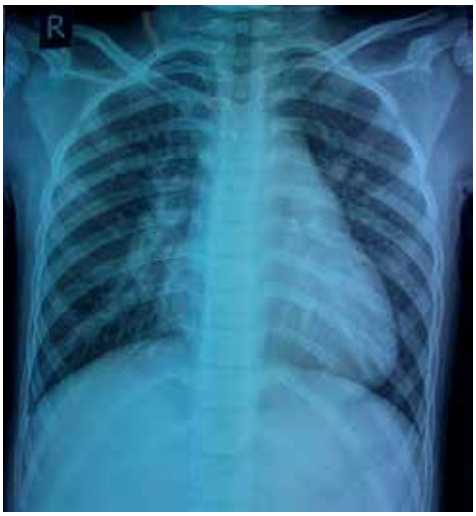

Fig. 4. Chest X-Ray P/A view showing cardiomegaly improvement with cardiothoracic ratio (CTR) 0.55

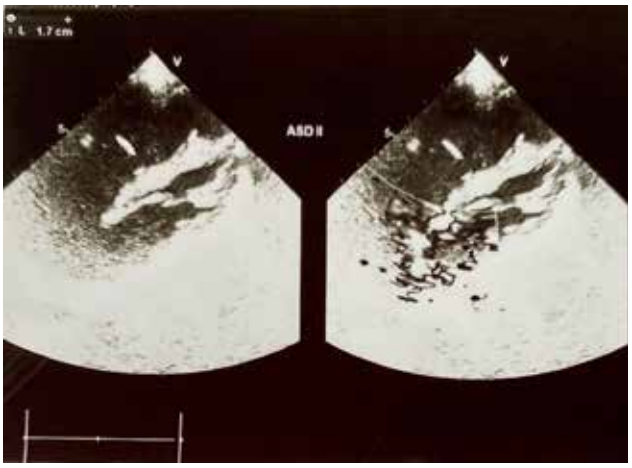

Fig. 5. Echocardiography showing the ostium secundum ASD (Diameter 1.7 cm)

and echocardiography examination were performed, a cardiomegaly and ASD, respectively were identified. The echocardiography revealed an ASD 1.88 cm with a left-to-right shunt (Figure 2). Echocardiography showed LVEF of 73.1%, and a sign of right atrium, and right ventricle (RV) dilatation (Figure 2, and 3), respectively. Based on this case, a diagnosed of Heart failure et causa ASD was made. The patient treatment was commenced on furosemide, and spironolactone. He was receiving furosemide 5 mg, and spironolactone 6,25 mg. After 3 months of evaluation, there is a clinical improvement and improved result of CXR, echocardiography and electrocardiography (ECG) (Figure 4, 5 and 6), respectively. The CXR shown CTR 0.55 (Figure 4). The echocardiography had shown that the hole of secundum ASD have gotten smaller than before (Figure 5). An ECG had shown a right ventricular hypertrophy (RVH), the heart rate of 89 bpm, PR interval of 176 ms, QRS duration of 120 ms, right axis deviation (Figure 6).

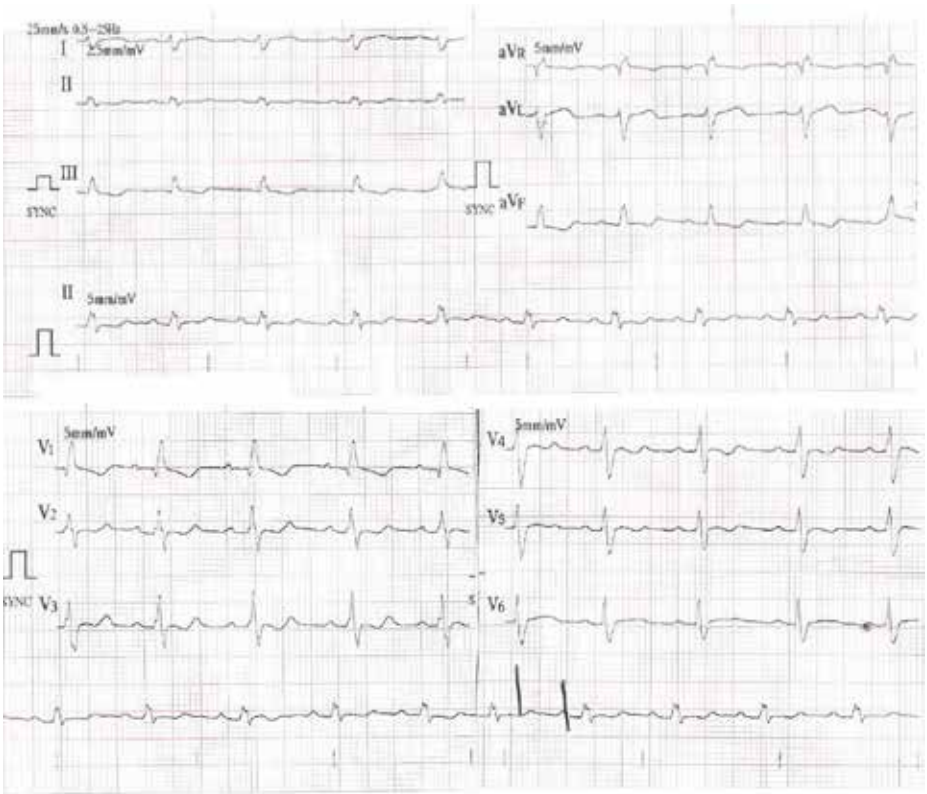

Fig. 6. Electrocardiography showing right ventricular hypertrophy (RVH)

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Discussions

This case describes a 12-year-old boy previous undetected ASD attended an ED due to sudden onset of dyspnoea that didn't improve at rest. Heart failure (HF) on paediatric is defined as a complex clinical syndrome that caused by cardiac structure or function abnormalities. It was an important thing that caused of morbidity and mortality not only in adult, but also in childhood [8]. Children whose history of HF have more than 20 times risk of death. The American Heart Association (AHA) estimated more than 8 million people in the United States (US) will be able to has HF, and the cost of treating patients whose HF around $160 billion on 2030 [9]. Impairing of ventricular filling or blood ejection fraction, or combination of both to systemic was important thing that can cause of HF [10].

Heart failure has been classified according to left ventricular ejection fraction (LVEF) (table 1). It is important to classify patients with HF because the difference prognosis and response to interventions. LVEF classified into 4 type: (1) heart failure with preserved ejection fraction (HFpEF, LVEF≥50 %); (2) heart failure with mildly reduced ejection fraction (HFmrEF, LVEF 41 %-49 %); (3) heart failure with improved ejection fraction (HFimpEF, previous LVEF ⩽ 40 % and a follow-up measurement LVEF > 40 %); and (4) heart failure with reduced ejection fraction (HFrEF, the LVEF is ⩽40 %) [11]. Children whose HF may have sign and symptom include recurrent dyspnoea, fatigue, growth failure, and exercise intolerance [8,12]. The NYHA is a subjective HF classification by a clinician used to characterized symptoms of patients with symptomatic or asymptomatic HF (table 2) [8]. The NYHA HF classification isn't easily applicable to younger children. Meanwhile, the modified ross classification is used for HF in younger children (table 3) [8, 13, 14].

The diagnose of HF was made based on framingham diagnostic criteria for heart failure. It must presence of 2 major or 1 major and 2 minor criteria were required to make the diagnosis of HF [15]. The criteria of framingham diagnosis are as follow: (1) major criteria; and (2) minor criteria. Major criteria consist of: acute pulmonary oedema; radiographic cardiomegaly; paroxysmal nocturnal dyspnoea; pulmonary rales; neck vein distension; hepatojugular reflex; third heart sound (S3 Gallop). Minor criteria consist of; ankle oedema; dyspnoea on exertion; hepatomegaly; pleural effusion; nocturnal cough; tachycardia (>120 beats per minute) [15].

Congenital heart disease (CHD) is one of predisposing factor that can be triggering HF on paediatrics [2, 3, 10, 16]. ASD is one of CHD that defined as a defect in the septum that divided atria of the heart, it was occurred at birth[16]. ASD cause left-to-right shunting that may result in right heart enlargement and right ventricle (RV) dysfunction. It will lead diastolic dysfunction [13]. The defect in cardiac septum

Table 1. **Classification of HF by LVEF [8]**

Type of HF According to LVEF	Criteria
HFrEF (HF with Reduced EF)	LVEF ⩽40 %
HFimpEF (HF with improved EF)	Previous LVEF ⩽40 % and a follow-up measurement of LVEF >40 %
HFmrEF (HF with mildly reduced EF)	LVEF 41–49 % Evidence of spontaneous of provokable increased LV filling pressures (eg, elevated natriuretic peptide, non-invasive and invasive hemodynamic measurement)
HFpEF (HF with preserved EF)	LVEF >50 %

Table 2. **Classification of HF by NYHA (13)**

	Class	Functional Capacity
NYHA Functional Classification System	I	Patients with heart disease but not causing physical activity restriction. Regular physical activity does not cause excessive fatigue, palpitation, dyspnea, or anginal pain.
	II	Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity result in fatigue, palpitation, dyspnoea, or anginal pain.
	III	Patients with heart disease who have limited physical activity. They feel comfortable at rest. Less activity than ordinary causes fatigue, palpitation, dyspnoea, or anginal pain.
	IV	Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Even while at rest, HF or the anginal syndrome symptoms may be present. If any physical activity is undertaken, discomfort increases.

Table 3. **Classification of Modified Ross (14)**

Modified Ross Classification for Paediatric Heart Failure	
Class I	Asymptomatic
Class II	Mild tachypnoea or diaphoresis with feeding in infants
Class III	Marked tachypnoea or diaphoresis with feeding in infants. Prolonged feeding times with growth failure Marked dyspnoea on exertion in older children
Class IV	Symptoms such as tachypnoea, retractions, grunting, or diaphoresis at rest

can also reduce stroke volume and would make inability to maintain cardiac output (CO). It will trigger sympathetic nervous system (SNS) and the renin-angiotensin-aldosterone system (RAAS) to be activated. The result of those are sodium and fluid retention occur in attempt to sustain preload and CO[17]. SNS, and RAAS were able to stimulate peripheral vascular vasoconstriction, so the blood pressure will increase to adequate organ perfusion. The effects of afterload changing and stress on blood vessels lead myocardial cell growth and adaptation. Those reaction will increase myocardial oxygen consumption, ventricular afterload which causes haemodynamic derangements such as abnormal pressure or volume loading, ventricular hypertrophy, myocardial ischemia, fibrosis interstitial, and decreases the density of cardiac capillaries. So that, it can cause cardiac toxicity and progression of HF [12]. HF in paediatrics can down-regulation both of β1 and β2-adrenergic receptors. In the Woulfe studied explained, that there was a difference occurrence of fibrosis and fibrotic gene expression in adult HF compared to paediatrics HF. Paediatrics HF showed less fibrosis and fibrotic gene expression than adult HF [18].

The clinical manifestation of HF in paediatrics whose CHD may vary significantly by their defect or age. Dyspnoea, fatigue, orthopnoea, fluid retention, falling asleep when feeding or becoming too tired to eat, malnutrition, failure to gain weight, and pulmonary & systemic congestion are clinical manifestations of HF in paediatrics, however each child may have experience symptoms differently [19]. Therefore, the application of general HF classifications in paediatrics such as the NYHA categories or the ross classification are important to provide a global assessment of HF severity in paediatrics [20].

The patient in this case presented with sudden onset of dyspnoea triggered by moderate exercise, which characterized him class II according to the NYHA, and Ross classification. Dyspnoea and fatigue can cause exercise intolerance among children with HF related to cardiac dysfunction. Exercise intolerance is a major cause of reduced quality of life in patients of HF. It could be happened because there was hypoperfusion of the lung, and skeletal muscular pathology significantly present in HF. Those mechanism leads to recurrent dyspnoea, as result of excessive activity more fully than before [20]. The patient has history of presence orthopnea, the presence of that has a positive predictive for HF. Orthopnea is the shortness of breath while supine brought on by an increase in the amount of blood flowing from the heart and pooling in the legs. While lying with prone position has an improve on orthopnea [21].

Making a diagnosis is a challenge when the clinical assessment does not correlate with clinical findings of investigation. Not only a comprehensive history, but also critical analysis of the symptom at the clinical findings are vital to make any diagnosis. The first step in diagnostic procedure in children of HF is based on non-invasive clinical investigations[13]. In this case, patient has a history of orthopnoea and radiographic cardiomegaly, so this patient have 2 major Framingham leading to HF. Further investigation is needed to determine the cause of HF. Echocardiography is the primary imaging modality in paediatric cardiology used for initial diagnosis especially in HF [12]. It provides an excellent image of cardiac structural and functional among children. In this case, the echocardiography showing the ostium secundum ASD (Figure 2). Patients with ostium secundum defect commonly asymptomatic during childhood. The diameter of defect smaller than 8 mm can be closed spontaneously, but the probability of spontaneous closure become poor if the diameter over than 10 mm. So that, it can cause a probability of HF, and the prevalence has been estimated around 10 %. In this case, the patient has diameter of defect 18.8 mm, it can increase the risk of HF occurrence.

The septum secundum is formed by the atrial roof to the right of the septum primum. As it is going to grow caudally and will cover the ostium secundum, the space between septum primum and septum secundum is known as the foramen ovale. The foramen ovale in the fetus allows blood that rich of oxygen pass the lung by flowing from right atrium to the left atrium. The Foramen is going to close spontaneously when the neonates was born and began to breathe. It caused by the change of pulmonary vascular resistance. So that, an ostium secundum ASD will cause left-to-right shunting that may result in chronic right ventricular volume overload, right heart enlargement and RV dysfunction. It will lead reverse-bernheim effect. This phenomenon was referred to characterized a syndrome of HF and usually present with cardiomegaly [3,22].

Cardiomegaly means enlargement of heart, it can happens due to ASD [23]. ASD has been found as-

sociated Gly247Asp ACTC1 mutation leading dilated hypertrophy, fibrosis, and contractile disfunction that cause dilatated cardiomyopathy (DCM) [24]. Chest X-ray (CXR) is one of basic investigation to detect cardiomegaly. Cardiomegaly in children suggested by the cardiothoracic ratio (CTR) > 55 %[25]. Therefore, the findings cardiomegaly in this case is associated with heart failure (Figure 1).

Medical therapy of HF in children focuses on 3 main goals: (1) reduce of pulmonary pressure; (2) increase of cardiac output & the improvement of organ perfusion; and (3) progression of HF is delayed [14]. The NYHA class of HF stage serves as the foundation for the management recommendations in the American College of Cardiology Foundation/American Heart Association guidelines [8]. Pharmaceutical treatments for HF patients with CHD aim to decrease pulmonary and systemic congestion by employing diuretics to increase contractility with inotropes and decrease afterload pressure. The other drug that routinely used in the pharmacological therapies of paediatric HF include angiotensin-converting enzyme inhibitors (ACEIs), spironolactone, and β-blockers [8,25].

Based on this case, patients used furosemide and spironolactone as his medical therapy. Furosemide is one of loop diuretics. The majority of HF patients should utilize diuretics instead. It will inhibit the reabsorption of sodium or chloride at the loop of henle, whereas spironolactone is one of aldosterone receptor antagonists (potassium sparing diuretics) act in the collecting duct [8,20]. The purpose of diuretic treatment is to reduce clinical manifestation of recurrent dyspnoea that caused by fluid retention using the lowest dose possible [8]. Tsujimoto studied showed spironolactone was an effective add-on therapy for patients with HF taking ACEIs, β-blockers, Calcium channel blockers, and diuretics. It blocks the RAAS pathway to prevent remodelling that will leading myocardial fibrosis [26].

Conclusions

This case described symptoms of HF due to CHD, particularly ASD. Currently, because of effective drug therapy, symptoms of HF due to ASD is able to reduce. However, it is still necessary to pay attention to the fact that heart failure in younger people can result from untreated CHD. In this case report, the patient had developed symptomatic HF with LVEF ⩾ 50 % also RA and RV dilatation. Most likely this patient developed HF due to the delayed diagnosis of ASD.

Conflicts of interest

The author declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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