CASE REPORT

Pediatrics Sciences Journal (*PedScij*) 2022, Volume 3, Number 2: 20-23
P-ISSN: 2722-0427, E-ISSN: 2722-1474

Pediatric Sciences Journal

Published by
Pediatrics Sciences Journal

Pericardial effusion in status epilepticus and global developmental delay one-year-old patient with co-exist of atrial septal defect: a case report

Pradhika Perdana Sakti[1*], Ariani[2]

[1]Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Brawijaya, Saiful Anwar General Hospital, Indonesia
[2]Department of Child Health, Faculty of Medicine, Universitas Brawijaya, Saiful Anwar General Hospital, Malang, Indonesia

*Corresponding to:
Pradhika Perdana Sakti. Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Brawijaya, Saiful Anwar General Hospital, Indonesia;
pradhikaperdana94@gmail.com

Received: 2022-08-17
Accepted: 2022-10-18
Published: 2022-12-14

ABSTRACT

Introduction: *Atrial septal* defect (ASD) is one of the most common types of congenital heart disease (CHD) and occurs due to spectrum closure failure during the intrauterine period. ASD is generally asymptomatic, but it can affect patients from various aspects.

Case presentation: A 1-year-old 4-month-old girl was referred with a major complaint of shortness of breath as early as 6 days before hospital admission. The dullness accompanied by a fever disappears, as convulsions, pallor and vomiting. On physical examination, a positive result in pathological reflex was found in a decrease in consciousness, epicantal fold, conjunctiva and pale skin, and edema of the left limb. The patient was examined for echocardiography and found the presence of moderate primum ASD with a left to the right pyre, mild tricuspid regurgitation with the possibility of pulmonary hypertension, and effusion pericardium with the collapse of the right atrium. In the laboratory results show the presence of normochromic normocytic anemia, leukocytosis, mild hyponatremia, hypokalemia, hypocalcemia, hypophosphatemia, increased PCT, increased SGOT / SGPT, PT elongation, a decreased of free T4 and increase in TSH, IgG CMV (+), IgG toxoplasma (+), IgG HSV-1 (+), IgG HSV-2 (+), and IgG Rubella (+). The patient is given treated at the PICU and provided with the help of ventilation, antibiotics, antipyretics, anticonvulsants, inotropic, and sympathomimetic agents. On the ninth day of care, the patient experienced cardiac arrest. RJP was performed, but the patient did not have ROSC and was pronounced dead.

Conclusion: ASD affects the patient's condition as well as the patient's survival both directly and indirectly. There is a link between ASD and developmental disorders, pericardial effusion and patient mortality. It is necessary to conduct a comprehensive evaluation and management of patients with ASD.

Keywords: Atrial septal defect, developmental disorders, encephalitis, pericarditis, pericardium effusion.
Cite This Article: Sakti, P.P., Ariani. 2022. Pericardial effusion in status epilepticus and global developmental delay one-year-old patient with co-exist of atrial septal defect: a case report. *Pediatrics Sciences Journal* 3(2): 20-23. DOI: 10.51559/pedscij.v3i2.33

INTRODUCTION

Congenital heart disease (CHD) is a condition in which anatomical malformations of the heart or large vessels occur before birth.[1] The prevalence of CHD globally in 2017 was 17.9/1000 and was the cause of 261,247 deaths.[2] CHD is broadly divided into cyanotic and asianotic heart diseases, of which *Atrial Septal Defect* (ASD) is one of them. ASD is a condition of failure to close the septum between the right and left atriums.[3]

ASD is one of the types of CHD that is widely found and associated with various syndromes, one of which is Down syndrome.[3,4] ASD patients are generally asymptomatic and, in small ASDs, are expected to have spontaneous closure without invasive measures.[3] However, ASD can impact various aspects and affect the patient's condition in general. Patients with CHD were found to have a higher risk of impaired neural and psychosocial development.[5,6] Patients with ASD are also susceptible to infection, especially patients who have experienced worsening in the form of Eisenmenger syndrome.[3] The risk of recurrent infection increases the risk of pericarditis, one of the most frequent etiologies.[7] The focus of primary infection can be on various organs, both in the respiratory system and the central nervous system (CNS).[8]

Pericarditis is one of the mechanisms of pericardium effusion. Pleural effusion is the accumulation of fluid in the cavity of the pericardium. Pericardial effusion generally does not cause symptoms but can also cause hemodynamic and life-threatening disorders like cardiac tamponades. Cardiac tamponade requires rapid management in fluid drainage through pericardiocentesis or surgery in several conditions.[7]

ASD can be seen as a type of CHD that does not interfere with the patient's life. However, there are many things to note regarding the impact of ASD on the patient. Patients need other treatments besides management for CHD and require close monitoring from an early age.[6] This report contains a case of a girl, 1 year and 4 months, with moderate ASD primum, who experienced several comorbidities and complications.

CASE PRESENTATION

A 1-year-4-month-old girl patient with *a primum atrial septal defect* (ASD) was referred from RSIA Amanah to the emergency installation (IGD) of Dr. Hospital. Saiful Anwar. The patient came with the main complaint of tightness from 6 days before entering the hospital. The patient also complained of a fever gone with the highest temperature of 38.5°C, which improved with the administration of paracetamol. There was a history of five seizures since 6 days before entering the hospital, with the characteristic eyes glancing upwards accompanied by the patient's hands and feet being stiff and limp repeatedly. The patient is unconscious after having a seizure. Seizures occur a maximum of 5 times a day. The patient was said to have looked pale since 7 days earlier, accompanied by vomiting 1 time containing approximately 120 ml of milk. There were no other complaints of coughing, runny nose, black bowel movements or vomiting blood.

The patient had a prior history of pneumonia and pericardial effusion 7 months before entering the hospital. Patients are also known to have a small primum ASD after echocardiography. In the history of pregnancy, pregnant women at the age of 35 years routinely control the midwife. During pregnancy, the patient's mother was once performed an ultrasonogram (US) examination by an obstetrics-gynecology specialist and was said to be normal. A history of taking medication routinely and rashes with fever in the first trimester are refuted. The patient was born in the hospital at 39-40 weeks gestation, with a history of early rupture of the amniotic (KPD) < 12 hours, birth weight 2900 grams and body length 48 cm. The patient did not cry immediately at birth and was found to be cramped. Patients have been given BCG, Pentabio, Measles and Hepatitis B0 immunizations.

On physical examination, the patient experienced a decrease in consciousness with a *Glasgow Coma Scale* (GCS) value of E3M2V4. Vital signs were found to be respiratory rate 35 times per minute, pulse 140 times per minute, and temperature 37.2°C. Patient weight 8.8 kg (P50-P75), height 77 cm (P90), head circumference 43 cm (P25-P50) and 16 cm upper arm circumference (P50). On examination of the localist status of the epicantal fold (+), the eyes and skin were found to be anemic without signs of icteric, palpable liver and lien, and there was edema in the left leg. Physiological reflexes include the biceps, tricep, Achilles and patella 2+/2+. Both sides found positive results when examining the pathological reflexes of Chadok, Gordon and Babinski.

Then the patient is carried out supporting examinations in the form of echocardiography and laboratory examinations. The echocardiography examination showed moderate primum ASD (7 mm) with a left to right shunt and mild tricuspid regurgitation with a high probability of pulmonary hypertension. Pericardial effusion was also found as much as 6 mm in the left posterior ventricle, 6 mm lateral left ventricle, and 7.7 mm superior right atrium with right atrial collapse.

Laboratory examination includes a complete blood test, clinical chemistry, immunoserological faal hemostasis, and body fluids. Results of laboratory examination: hemoglobin 9.10 g/dL, erythroid 3.16 million, leukocytes 12.37 $10^3/mm^3$, hematocrit 26.40%, platelets 194 $10^3/mm^3$, MCV 83.50 $\mu m10^3$, MCH 28.80 pg, MCHC 34.50 g/ dL, RDW 14.50%, PDW 9.0 fL, MPV 9.1 fL, P-LCR 16.6%, PCT 0.18%, NRBC absolute 0.01 $10^3/\mu L$, percent NRBC 0.1%. On the calculation of the types found: eosinophils 0.00%, basophils 0.10%, neutrophils 84.70%, lymphocytes 10.00%, monocytes 5.20%, absolute eosinophils 0.00 $10^3/mm^3$, absolute basophils 0.01 $10^3/mm^3$, absolute lymphocytes 1.24 $10^3/mm^3$, NLR 8.45, absolute monocytes 0.64 $10^3/mm^3$, immature granulocytes 0.09 $10^3/\mu L$ (0.7%). The patient is also subjected to a clinical chemical examination. In faal hemostasis: PATIENT PT 26.70 seconds (control 11.9 seconds), INR 2.69, APTT patient 29.40 seconds (control 25.2 seconds). On examination of the liver faal: SGOT 699 U /L. Blood gas analysis test results: pH 7.37, pCO2 27.9 mmHg, pO2 103.0 mmHg, HCO3 16.3, BE -9.2 mmol/L, O2 saturation 97.5% and Hb 9.7 g/dL. Lactic acid examination showed a 2.6 mmol/L. Renal faal results: ureum 51.9 mg/dL, and creatinine 0.51. On electrolyte examination: Sodium 135 mmol/L, potassium 3.14 mmol/L, chloride 104 mmol/L. In immunoserology, thyroid test results: Free T4 0.40 ng/dL, TSH 6.51 µU/mL, and procalcitonin 3.71 ng/mL. In addition, IgG toxoplasma, IgG rubella, IgG anti-HSV-2, IgG anti-HSV-1 and IgG anti-CMV reactive were found. The patient has carried out an examination of body fluids in the form of LCS fluid analysis. Chemical test results: protein 13.8 mg/dL, glucose 86 mg / dL, LDH 19 IU / L. Macroscopic examination results: colorless, clear, and clot (-). Microscopic examination results: no erythrocytes, leukocytes, PMN or MN cells were found. On special tests were found Nonne (-) and Pandy (-).

Patients diagnosed with epilepticus status et cause prolonged hypoxia dd encephalopathy dd encephalitis, respiratory failure et cause pneumonia, pneumonia, suspect Down syndrome, distributive shock et causa hypoxia, congenital hypothyroidism on treatment and *global developmental delay*, minimal pericardial effusion. The patient was then given an intravenous injection of ceftriaxone 2x450 m, paracetamol 3x90 mg, phenytoin 2x16 mg, dobutamine 12.5 mg/kgBB/min and epinephrine 0.05 mg/kgBB/min. Breathing assistance is also provided in *noninvasive ventilation–spontaneous / time* (NIV-ST) PEEP 7 cmH_2O Psupp 12 cm H2O, FiO_2 45%. Furthermore, the patient is treated at the *Paediatric Intensive Care Unit* (PICU), and intubation is carried out.

During the patient's treatment, periodic evaluations and adjustments to the management are carried out. On the ninth day of treatment, the patient experienced desaturation with 36% SaO2, followed by cardiac arrest and respiratory arrest. Patients were given cardiopulmonary resuscitation (RJP) 5 cycles and epinephrine intravenously 0.01 mg - 0.03 mg/kgBB every 3-5 minutes. The patient did not experience a *return of spontaneous circulation* (ROSC) and was found to be maximum pupil mydriasis. The patient was later pronounced dead.

DISCUSSION

Congenital heart disease (CHD) is an anatomical malformation in the heart or large vessels that occurs during intrauterine development.[1] The prevalence of CHD worldwide is 17.9/1000, with 19.1/1000 men and 16.6/1000 women.[4] Based on global data in 2017, CHD was

the cause of 261,247 deaths, with 180,624 deaths occurring in children less than one-year-old. Deaths from CHD are highest in the lower and lower socio-demographic indices.[2]

CHD is divided into two, namely cyanotic and asianotic heart disease. *Atrial Septal Defect* (ASD) belongs to the asianotic group. ASD occurs due to the failure of communication closure between the right and left atria.[3] Based on the location of the defect, ASD can be divided into Secundum ASD, primum ASD, venosus sinus ASD, and other rarely found types of ASD. The Secundum ASD defect, the most common type, is located in the central part of the primum septum that develops from the middle and reef to the lower atrial cavity to meet with endocardial bearings. Primum ASD occurs due to a deficiency of endocardial bearing proliferation. ASD sinus venosus is characterized by defects near the superior vena cava and the upper right pulmonary vein.[9]

ASD, along with *Ventricular Septal Defect* (VSD), dominates the incidence of CHD with an incidence percentage of 29.6% of all CHD cases.[4] ASD is also found in many patients with the syndrome, one of which is Down syndrome.[3] Generally, ASD patients have no symptoms or are asymptomatic. Patients with ASD less than 5 mm can be completely asymptomatic. While in patients with ASD 5-10 mm, symptoms are found at the age of four or five decades. If the ASD is larger, symptoms are found in the third decade of life. Symptoms can be dyspnea, fatigue, intolerance of physical activity, palpitations or a sign of right heart failure. In physical examination, a systolic ejection murmur can be found in the pulmonary area accompanied by *wide fixed splitting* S2. Late or inadequately treated ASD can lead to several complications, such as atrial dysrhythmia, pulmonary arterial hypertension, congestive right heart failure, *transient ischemic attack* (TIA), stroke, and Eisenmenger syndrome.[3]

CHD was also found to often impact other aspects, such as developmental disorders and infections.[6] CHD was associated with an increased risk of neural and psychosocial development morbidity. Developmental delays and disabilities are the most common long-term morbidities in children with CHD.[5,6] Children with 14 times higher CHD experienced impaired physical limitations such as crawling, walking and running.[6] The output of these comorbidities is worse in patients with social risk factors such as racial minorities, lack of private insurance, and low levels of maternal education. Such factors are generally unmodifiable and affect the first two to three years of a patient's life. This indicates that CHD affects many aspects of the patient's life. Regular developmental assessments must be carried out to assess the presence or absence of impairment.[1] Patient needs additional care beyond the treatment of handling the direct effects of CHD and requires close monitoring from an early age.[6]

Infection is also one of the things that are often found in children with CHD. A cohort study in Indonesia by Djer et al. found that children with non-cyanotic CHDs of left-to-right shunt had almost twice the risk of developing tract infections acute respiratory (ARI) and a longer risk of recurrence and duration than children without CHD.[10] There is an increased relative risk of hospitalization in children with CHD affected by ARI. The risk is higher in children with severe CHD.[11] In children with ASD who have replicated Eisenmenger syndrome, an increase in susceptibility to infection was found.[3] In this patient, there are signs that the patient has an infection, both currently ongoing infections such as encephalitis and pneumonia, as well as those that have been sufferers seen in immunoserological examinations in the form of IgG toxoplasma, rubella, anti-HSV-2, anti-HSV-1 and anti-CMV reactive.

The presence of infection in patients with ASD can be related to pericardium effusion with a possible etiology in the form of infection. Pericardial effusion can be caused by all diseases that give rise to pericarditis or involve the pericardium. Another mechanism is a decrease in fluid reabsorption.[7] One of the etiologies is infection by viruses, bacteria, fungi, protozoa and HIV-related. Clinical patients with pericardium effusion due to the focus infection are generally critical, and clinical infection is more dominant than clinical due to pericardium effusion.[12] In addition to ARI, CHD is associated with an increased risk of central nervous system (CNS) infection. Although the mechanism is not yet clear, it is estimated that the potential etiological factors are a decrease in the immune response, genetic abnormalities, abnormal physiological, medical exposure complex, and acquired infectious diseases such as endocarditis.[8] In these patients, ASD can cause pericardium effusion with increased patient responsiveness to infection. The existence of an etiology in the form of foci of infection in the lungs (pneumonia) and brain (encephalitis) in these patients increase the patient's risk of developing pericarditis and then progressing to pericardium effusion. In addition to being the etiology of pericarditis, the presence of encephalitis in patients also aggravates the work of the heart. Encephalitis can replicate in the heart, causing *ballooning* of the apex, acute heart failure, and sinus arrest.[13–15] In addition, brain or CNS injuries can impact the heart through *neurogenic stunned myocardium* (NSM) syndrome or *neurogenic stress cardiomyopathy* (NSC). NSC can occur due to *subarachnoid hemorrhage* (SAH), brain trauma, stroke, acute stress, epileptiform seizures or infections in the CNS.[15,16] The pathophysiology involved is the release of excess catecholamines during neurological needs that lead to increased need, stress, and heart myositis.[17]

Pericarditis in adult patients, based on the 2015 ESC guidelines, is enforced in the presence of 2 of 4 signs: (1) chest pain pericarditis; (2) *pericardial rub*; (3) on the ECG found st elevation or new PR depression; (4) pleural effusion (new or old). In addition, using imaging techniques, supporting findings can be found in the form of increased inflammatory markers and evidence of inflammation of the pericardium. However, there are no criteria for diagnosing pericarditis in a child yet. Inflammation of the pericardium, such as pericarditis, or decreased reabsorption of pericardial fluid due to a general increase in systemic venous pressure due to congestive heart failure or pulmonary hypertension can cause fluid discharge or pericardium effusion.[7] In patients, pericardial effusion is suspected to be caused by pericarditis characterized by an increase in inflammatory markers, procalcitonin, by 3.71 ng/mL. From the echocardiography results, the patient is also said to have the possibility of

experiencing pulmonary hypertension, which can cause an increase in systemic venous pressure, which then decreases the reabsorption of fluid in the pericardium and causes fluid accumulation.

Pericardial effusions can be divided by onset (acute, sub-acute and chronic), distribution (circumferential or localized), hemodynamic impact (absent, cardiac tamponade, effusive-constrictive), composition (exudate, transudate, blood, sometimes air or gas from a bacterial infection) and size. Sizes can be seen from the assessment of the echocardiogram and are divided into small (<10 mm), medium (10-20 mm) or large (>20 mm). The management of pericardial effusion is focused on its etiology. If effusion occurs due to pericarditis, then the management carried out follows the management of pericarditis, such as the administration of non-steroidal anti-inflammatory drugs (NSAIDs). If the patient begins to experience symptoms or fails initial management, then drainage of effusions with pericardiocentesis needs to be considered.[7] In patients, the total volume of pericardium outflow is 10-20 mm, which is said to be moderate pericardium effusion. The patient is given antibiotics to cope with the etiology of the infection.

Patients who experience a slow accumulation of pericardial fluid usually do not experience symptoms. However, complaints and hemodynamic disorders will appear if fluid accumulation occurs suddenly and quickly. An emergency that can occur is cardiac tamponade, a life-threatening condition due to compression of the heart by the accumulation of pericardiac effusion sooner or later.[7] The liquid accumulating in the pericardium, if it continues to grow, will reach the point where the *pressure-volume relation* (PVR) decreases so that even the slightest increase in volume will cause an increase in intra-pericardial pressure and leads to cardiac tamponade.[12] When the intra-pericardial pressure exceeds the right atrial pressure, the right atrium collapses.[18] Atrial collapse is usually found before the ventricular collapse and results from progressive cardiac tamponade.[19] Cardiac tamponade management in the form of drainage of pericardial fluid, mainly through pericardiocentesis.[7] The patient, in this case, is presumed to have died from cardiac tamponade due to rapidly progressing pericardium effusion.

CONCLUSION

This report presents a case of a pediatric patient with *atrial septal* defect (ASD) who experienced various comorbidities and complications. ASD, a type of ASIANOTIC PJB for congenital heart disease (CHD), generally does not cause symptoms. ASD directly or indirectly affects the patient's condition and survival. The relationship between ASD illustrates this and developmental disorders, pericardial effusion and death of patients. Patients with pericardium effusion can be arranged according to the etiology and considered pericardiocentesis, especially in an emergency such as cardiac tamponade. This case raises awareness of the need for comprehensive evaluation and management of patients with ASD.

DISCLOSURE

Consent for Publication

Written informed consent was obtained from the patient to publish this case report.

Fundings

No fundings

Conflicts of interest

The author declared that there is no conflict of interest

Author contribution

All authors contributed equally in preparing and writing this editorial.

REFERENCE

1. Rao PS. Congenital Heart Defects – A Review. In: Rao PS, editor. Congenital Heart Defects-A Review. London: IntechOpen; 2012. Available from: ttps://www.intechopen.com/books/955
2. Zimmerman MS, Smith AGC, Sable CA, Echko MM, Wilner LB, Olsen HE, et al. Global, regional, and national burden of congenital heart disease, 1990–2017: a systematic analysis for the Global Burden of Disease Study 2017. Lancet Child Adolesc Health. 2020;4:185–200.
3. Menillo AM, Lee LS, Pearson-Shaver AL. Atrial Septal Defect. In: StatPearls. 2021. p. 1–10. Available from: https://www.ncbi.nlm.nih.gov/books/NBK535440/
4. Wu W, He J, Shao X. Incidence and mortality trend of congenital heart disease at the global, regional, and national level, 1990-2017. Vol. 99, Medicine. Lippincott Williams and Wilkins; 2020. p. 1–8.
5. Mussatto KA, Hoffmann R, Hoffman G, Tweddell JS, Bear L, Cao Y, et al. Risk factors for abnormal developmental trajectories in young children with congenital heart disease. Circulation. 2015;755–61.
6. Razzaghi H, Oster M, Reefhuis J. Long-term outcomes in children with congenital heart disease: National Health Interview Survey. Journal of Pediatrics. 2015;166(1):119-124.e1.
7. The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC). 2015 ESC Guidelines for the diagnosis and management of pericardial diseases. Vol. 36, European Heart Journal. Oxford University Press; 2015. p. 2921–64.
8. Bagge CN, Smit J, Madsen NL, Olsen M. Congenital Heart Disease and Risk of Central Nervous System Infections: A Nationwide Cohort Study. Pediatric Cardiol. 2020;869–76.
9. Puri K, Allen HD, Qureshi AM. Congenital Heart Disease. Pediatrics in Review. 2017;38(10):471–86. Available from: http://pedsinreview.aappublications.org/
10. Djer MM, Osmardin E, Hegar B, Setyanto DB. Increased Risk of Recurrent Acute Respiratory Infections in Children with Congenital Heart Disease: A Prospective Cohort Study. Indones Biomed J. 2020;12(4):355–60.
11. Granbom E, Fernlund E, Sunnegårdh J, Lundell B, Naumburg E. Respiratory Tract Infection and Risk of Hospitalization in Children with Congenital Heart Defects During Season and Off-Season: A Swedish National Study. Pediatric Cardiol. 2016;37:1098–105.
12. Azarbal A, LeWinter MM. Pericardial Effusion. Vol. 35, Cardiol Clin. W.B. Saunders; 2017. p. 515–24.
13. Braiman D, Constantino Y, Westreich R. When the brain slows the heart - Herpes encephalitis and sinus arrest: A case report. European Heart Journal - Case Reports. 2021;5(8):1–5.
14. Saini A, Patel B. Bubbles in the pericardium. Am J Case Rep. 2015;16:420–3.
15. Gelow J, Kruer M, Yadav V, Kaul S. Apical Ballooning Resulting from Limbic Encephalitis. The American Journal of Medicine. 2009;122(6):583–6.
16. Gopinath R, Ayya S. Neurogenic stress cardiomyopathy: What do we need to know. Annals of Cardiac Anaesthesia. 2018;21(3):228–34.
17. Biso S, Wongrakpanich S, Agrawal A, Yadlapati S, Kishlyansky M, Figueredo V. A Review of Neurogenic Stunned Myocardium. Cardiovascular Psychiatry and Neurology. Hindawi Limited; 2017. p. 1–6.
18. Argulian E, Ramirez R. Understanding Right Atrial Collapse: Timing Is Everything. Annals of Emergency Medicine. 2018;1–3.
19. Pérez-Casares A, Cesar S, Brunet-Garcia L, Sanchez-de-Toledo J. Echocardiographic evaluation of pericardial effusion and cardiac tamponade. Vol. 5, Frontiers in Pediatrics. Frontiers Media S.A.; 2017. p. 1–10.