



INTISARI SAINS MEDIS

Published by Intisari Sains Medis

## A 6-month-old patient with cleft lip palate, atrial septal defect, and presenting hyperkalemia: a case report



CrossMark

Ni Putu Anggita Medyantari<sup>1\*</sup>, Ayu Pisita Wulandari<sup>2</sup>, Zahra Fadhilzka Tiara<sup>3</sup>

<sup>1</sup>General Practitioner, Puri Bunda Mother and Child Hospital, Denpasar, Indonesia

<sup>2</sup>General Practitioner, Pelengkap Hospital, Jombang, Indonesia

<sup>3</sup>General Practitioner, Pondok Jati Mother and Child Hospital, Sidoarjo, Indonesia

\*Corresponding to:

Ni Putu Anggita Medyantari; General Practitioner, Puri Bunda Mother and Child Hospital, Denpasar, Indonesia; medyantari15@gmail.com

Received: 2024-02-04

Accepted: 2024-03-19

Published: 2024-04-22

### ABSTRACT

**Introduction:** Cleft Lip Palate (CLP) is a prevalent congenital anomaly in Indonesia. Gradual surgery is the gold standard treatment for the condition. However, screening for other comorbidities must be conducted before the surgery. Congenital heart Defects (CHDs) are one of the prevalent concurrent conditions of CLP patients. One of the CHD that can be found is Atrial Septal Defect (ASD). Moreover, electrolyte levels must also be assessed and treated if abnormal before surgery. Hyperkalemia is a type of electrolyte imbalance that can be found, possibly caused by increased potassium load or decreased excretion.

**Case description:** A 6-month-old girl was admitted to our hospital with a fever and dehydration. She complained of persistent vomiting. She had a history

of CLP and CHD since birth. The echocardiogram revealed Small Secundum ASD. Laboratory results showed an elevated potassium (5.52 mEq/l). The patient was planned to get primary cleft lip repair and tip rhinoplasty. The surgery was postponed due to the patient's condition and laboratory findings. During hospitalization, she received electrolyte imbalance correction and symptomatic treatment. A 1-week follow-up assessment showed stable condition, and the patient continued to prepare for the surgery.

**Conclusion:** ASD is frequently observed as one of the most prevalent comorbidities of a CLP patient. Dehydration can increase the potassium level of the blood, and it must be treated before the surgical procedure for CLP patients with ASD condition.

**Keywords:** atrial septal defect, cleft lip palate, hyperkalemia.

**Cite This Article:** Medyantari, N.P.A., Wulandari, A.P., Tiara, Z.F. 2024. A 6-month-old patient with cleft lip palate, atrial septal defect, and presenting hyperkalemia: a case report. *Intisari Sains Medis* 15(1): 459-462. DOI: 10.15562/ism.v15i1.1999

### INTRODUCTION

Cleft Lip Palate (CLP) is a congenital anomaly characterized by an abnormal upper lip, alveolus, or palate gap. This condition occurs due to the incomplete fusion of the frontonasal and maxillary processes and the palatal shelves of the maxillary processes during embryonic development.<sup>1</sup> CLP is associated with genetic mutation and maternal exposure to tobacco, alcohol, teratogenic medication, and vitamin deficiencies.<sup>2</sup> In Indonesia, CLP is one of the most prevalent congenital anomalies, with a national prevalence rate of 0.2%. The gold standard treatment for CLP is gradual surgery involving multidisciplinary doctors.<sup>3</sup>

Other comorbidities like anemia, respiratory problems, and cardiovascular

diseases, often complicate CLP conditions.<sup>2</sup> Congenital Heart Disease (CHD) is one of the most common comorbidities of CLP, with the prevalence ranging from 5.4 to 51 percent worldwide. The two conditions are related because the heart and palate develop during the 5<sup>th</sup> to 9<sup>th</sup> week of gestation.<sup>4</sup>

Atrial Septal Defect (ASD) is the most common type of CHD that occurs in CLP patients.<sup>4</sup> ASD is defined as a heart anomaly where there is an abnormal hole between the right and left atria of the heart. There are five types of ASD, such as: Patent foramen ovale, Ostium secundum defect, Ostium primum defect, Sinus venosus defect, Coronary sinus defect.<sup>5</sup>

Individuals with a minor ASD typically do not exhibit any symptoms. In contrast,

a more significant defect may result in symptoms such as shortness of breath, tiredness, difficulty exercising, irregular heartbeat, or indications of right-sided heart failure. The murmur can be assessed, featuring a soft, systolic ejection murmur heard in the second intercostal space, particularly in the pulmonic area, along with a wide, fixed splitting of the second heart sound (S2). Transthoracic echocardiography is the gold standard imaging modality, where a physician can examine the heart structure and function, measure the size of the defect, understand the blood flow direction and ratio, and detect other abnormalities of the heart. Treatment for ASD depends on the size of the defect. ASD with a diameter less than 5 mm usually closes spontaneously in

the first year of life, while a defect larger than 1 cm may require medical/surgical intervention.<sup>5</sup>

Before surgery, CLP patients must do several laboratory examinations, including complete blood count, electrolyte level, and other related examinations, since laboratory abnormalities may cause complications during an anesthetic procedure. One of the abnormalities that can be found is electrolyte imbalance, one of which is hyperkalemia. Hyperkalemia is when the potassium level is above 5,5 mEq/L. It can be caused by pseudohyperkalemia (commonly caused by an error in blood sampling), increased efflux from intracellular fluid (ex, acidosis, etc), increased potassium load (ex, high potassium diet, hemolysis, etc), decreased excretion (ex: acute renal injury, medication, Addison's disease, dehydration).<sup>6</sup>

Management for hyperkalemia depends on the potassium level and clinical manifestation. Possible causes must be identified and eliminated, complications must be treated, and potassium levels must be corrected without overcorrecting. There are several methods to treat hyperkalemia, such as calcium supplementation, pharmacotherapy to shift potassium from extracellular to intracellular fluid compartments, pharmacotherapy is a procedure to remove potassium excess (e.g., dialysis or diuretics).<sup>6</sup>

In the present case, a 6-month-old patient with CLP and ASD was presented, complicated by hyperkalemia found before surgery.

## CASE DESCRIPTION

A 6-month-old girl was admitted to our hospital with a chief complaint of fever. Fever occurred 3 days before admission. The parents said their children have persistent vomiting. It occurred one day before admission—frequency of vomiting more than 10 times. So, the intake was poor. Their last urination was 8 hours ago with a small volume. No diarrhea occurred before admission. From the history of medication, the patient was given paracetamol one hour before admission. There is no medication for vomiting. The patient had a history of CLP and CHD since birth. The echocardiogram revealed

Small Secundum ASD.

The patient's physical examination was somnolence, pulse was 108 times/minute, respiratory rate was 30 times/minute, and body temperature was 38°C. The patient's head and face examination were abnormal; the eyes were sunken, the sclera were anicteric, and the conjunctiva was not pale. The ear, nose, and throat examinations were normal. There was a CLP, and the patient had routine control with a plastic surgeon. No lymph node enlargement was found on the neck. The chest was regular and symmetrical on rest and movement; breath sounds were bronchovesicular without rales or wheezing; heart sounds were abnormal, and there was a murmur in auscultation. The abdomen was not distended, the liver and spleen were not palpable. The superior and inferior extremities were clammy, and the capillary refill time was 4. All the physical examinations showed signs of dehydration.

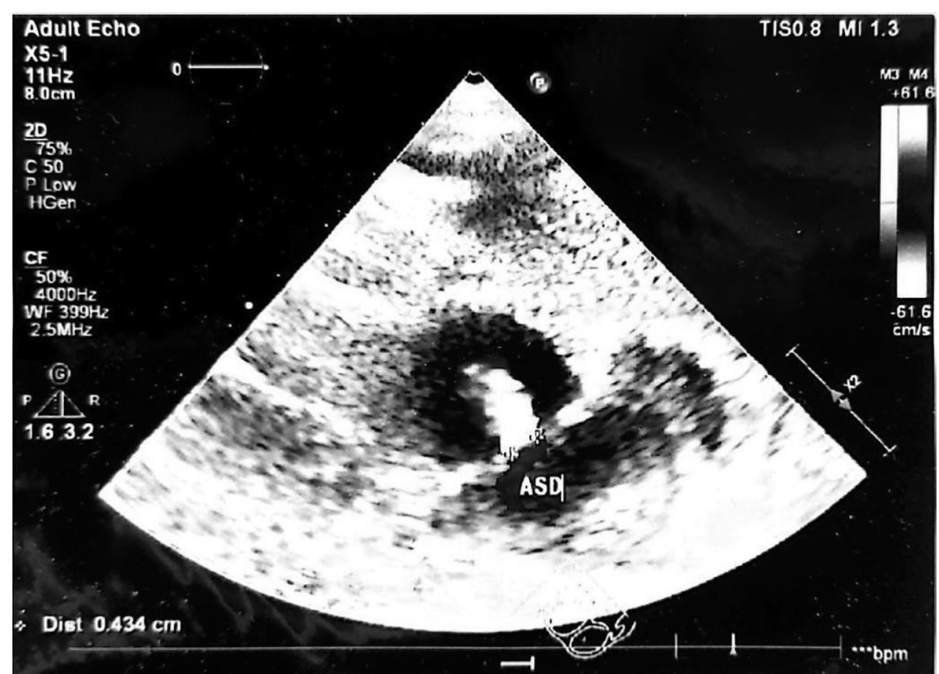
Laboratory results showed a white blood cell count of 9,830/uL, a hemoglobin level of 11.6 gr/dL, a platelet count of 395,000, and Neutrofil absolut 2530/uL. The patient also showed normal coagulation function at PPT level 11 seconds and APTT level 11 seconds. The renal function test of the patient is within normal limits; creatinine serum was 0,68 mg/dL, and the urea was

22,8 mg/dL. The OT slightly increased at 59 U/l, but the PT level was normal at 27 U/l. The electrolyte test showed an elevated potassium (5.52 mEq/l). The natrium level was normal (145 m3q/l).

The patient was planned to get primary cleft lip repair and tip rhinoplasty. The surgery was postponed due to the patient's condition and laboratory findings. During hospitalization, she received rehydration with intravenous fluids, electrolyte imbalance correction, and symptomatic treatment for the fever and vomiting by intravenous. The patient was discharged on the 4<sup>th</sup> day of hospitalization since the clinical condition and oral intake improved. A 1-week follow-up assessment showed stable condition, and the patient continued to prepare for the surgery.

## DISCUSSION

The case showed a patient with CLP and small secundum ASD, a type of CHD. CHD is one of the most common comorbidities of CLP, which may occur because both the heart and palate develop during the 5<sup>th</sup> to 9<sup>th</sup> week of gestation. Therefore, disruption of development during this age may cause congenital abnormalities in both organs.<sup>4</sup> ASD itself is one of the most common types of CHD that can be found in CLP patients.<sup>4</sup>



**Figure 1.** Small Secundum ASD sized 0.23 – 0.4 cm (Echocardiogram)

There are several types of ASD, one of which is secundum ASD, which occurred in this case. A secundum ASD is a flaw that develops in the fossa ovalis, usually caused by defects within the septum primum. It can lead to an atrial-level left-to-right shunt from late ventricular systole to early diastole. The dimension of secundum ASD ranges from a few millimeters to 3 centimeters. Defects smaller than 1 cm usually have less complications, such as a smaller shunt and minimal dilatation of the right heart chamber.<sup>7</sup>

The patient experienced no symptoms, consistent with most ASD cases, which remain asymptomatic during childhood. In contrast, adults with secundum ASD may experience degradation of exercise capacity and maximum oxygen uptake. Abnormal heart sounds may be found on physical examination, such as wide splitting of second heart sound, soft systolic ejection murmur over the pulmonary area in the left upper sternal border, diastolic rumble over the left lower sternum, and/or holosystolic murmur over the apex. Transthoracic echocardiography serves as the primary diagnostic tool for the condition. Through this examination, healthcare professionals can ascertain the presence, location, size, and hemodynamic characteristics of the ASD.<sup>7</sup>

This patient received no treatment for her ASD since the lesion found has a small size and no significant hemodynamic changes. This patient was scheduled to have follow-up echocardiography three months after the previous one. Closure of ASD itself is indicated where a hemodynamically significant shunt that causes right heart structure enlargement is found, proven by a pulmonary-to-systemic flow ratio greater than 1.5. Other indications are suspicion of paradoxical embolism and/or documented orthodeoxia-platypnoea. Patients with small ASD that do not have indications for surgical treatment should be followed up expectantly for the possibility of shunt increase later in life.<sup>7</sup>

The research on the correlation between kidney and cardiovascular diseases continues to grow. Researchers have made progress in understanding the various neurological, hormonal, and pathways of hemodynamics involved in cardiorenal diseases. In an initial

investigation using a case-control design conducted by Fuhrman et al. in 2019 at the Adult Congenital Heart Disease Center of Children's Hospital of Pittsburgh, researchers compared the baseline levels of biomarkers in the tubular system were assessed in a group of 30 young adults with congenital heart disease (CHD) to those of 30 healthy young adults. The study aimed to identify early signs of kidney injury before they become clinically evident. Neither group exhibited significant proteinuria or albuminuria. The CHD group displayed normal kidney function, as indicated by serum creatinine and estimated glomerular filtration rate (eGFR), and none of the CHD patients had documented hypoxemia, which is known to affect renal function negatively. Results indicated that compared to those without CHD, patients with CHD showed notably higher levels of kidney injury molecule-1/creatinine (KIM-1/Cr) in their urine and lower concentrations of tissue inhibitor of metalloproteinases-2 (TIMP-2) and insulin-like growth factor binding protein 7 (IGFBP7). These biomarkers could potentially be used to detect early tubular injury before abnormalities in traditional biomarkers of kidney dysfunction are apparent.<sup>8</sup>

According to research by Madsen et al. (2017), AKI in patients with congenital heart disease is common and associated with an increased risk for CKD.<sup>9</sup> Acute kidney injury involves a sudden decrease in kidney function, spanning from slight impairment to complete renal failure necessitating dialysis. The condition is currently defined using standardized criteria, notably the Kidney Disease Improving Global Outcomes (KDIGO) criteria.<sup>10</sup> Unfortunately, very few reports were found that discussed CLP with CHD.

## CONCLUSION

We reported a 6-month-old girl was admitted to our hospital with a chief complaint of fever. Fever occurred 3 days before admission and persistent vomiting. All the physical examinations showed signs of dehydration. The electrolyte test showed an elevated potassium (5.52 mEq/l). The patient was planned to get primary cleft lip repair and tip rhinoplasty.

The surgery was postponed due to the patient's condition and laboratory findings. During hospitalization, she received rehydration with intravenous fluids, electrolyte imbalance correction, and symptomatic treatment for the fever and vomiting by intravenous. A 1-week follow-up assessment showed stable condition, and the patient continued to prepare for the surgery.

ASD is one of the most common comorbidities of a CLP patient. Dehydration can increase the potassium level of the blood, and it must be treated before the surgical procedure for CLP patients with ASD condition.

## DISCLOSURE

### Conflict of Interest

All authors stated that there is no conflict of interest.

### Funding

None.

### Author Contribution

All.

### Ethical Consideration

This publication has been permitted to publish the patient data by the patient's parents through informed consent.

## REFERENCES

1. Chaudary S. Cleft Lip and Palate – A Review Article. *International Journal of Research and Review*, 2022; 8(7):236-243.
2. Silva HPV, Arruda TTS, Souza KSC, Bezerra JF, Leite GCP, et al. Risk factors and comorbidities in Brazilian patients with orofacial clefts. *Original Research Community Dental Health*. 2018; 32(24):1-12.
3. Moelek NF. Pedomon Nasional Pelayanan Kedokteran Tata Laksana Bibir Sumbing dan Lelangit. Kementerian Kesehatan Indonesia. 2019. Tersedia di: [https://yankes.kemkes.go.id/unduh/fileunduh\\_1660185991\\_70505.pdf](https://yankes.kemkes.go.id/unduh/fileunduh_1660185991_70505.pdf)
4. Akhiwu BI, Efunkeya AA, Akhiwu HO, Adebola RA. Congenital Heart Disease in Cleft Lip and Palate Patients : How Common is The Association?. *Journal of Advanced Oral Research*, 2017; 8(1&2): 53-56.
5. Menillo AM, Lee LS, Pearson-Shaver AL. 2022. Tersedia di: <https://www.ncbi.nlm.nih.gov/books/NBK535440/>
6. Daly K, Farrington E. 2013. Hypokalemia and Hyperkalemia in Infants and Children : Pathophysiology and Treatment. *Journal of Pediatric Health Care*. 2013;27(6):486-496.

7. Geva T, Martins JD, Wald RM. Atrial septal defects. *Lancet*. 2014;383:1921-1932.
8. Fuhrman DY, Nguyen L, Hinds M, Kellum JA. Baseline tubular biomarkers in young adults with congenital heart disease as compared to healthy young adults: Detecting subclinical kidney injury. *Congenit Heart Dis*. 2019;14(6):963-967. doi: [10.1111/chd.12862](https://doi.org/10.1111/chd.12862)
9. Madsen NL, Goldstein SL, Frølev T, Christiansen CF, Olsen M. Cardiac surgery in patients with congenital heart disease is associated with acute kidney injury and the risk of chronic kidney disease. *Kidney Int*. 2017;92(3):751-756. doi: [10.1016/j.kint.2017.02.021](https://doi.org/10.1016/j.kint.2017.02.021)
10. KDIGO Clinical Practice Guideline for Acute Kidney Injury. *Kidney International Supplements*. 2012, 2(1):1-141. doi: [10.1038/kisup.2012.3](https://doi.org/10.1038/kisup.2012.3)



This work is licensed under a Creative Commons Attribution