


Hypoxic Spell in an Adult

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ABSTRACT

Introduction: Hypoxic spells are serious and potentially fatal complications of cyanotic congenital heart disease. These events typically occur in children and are extremely rare in adults. They are caused by a sudden reduction in pulmonary blood flow, leading to severe hypoxia, cyanosis, seizures, and even death. Although rare, hypoxic spells should be considered as a differential diagnosis in adult patients presenting with hypercyanotic crises, particularly those without a known history of congenital heart disease. **Case**: A 20-year-old woman presented with worsening shortness of breath and cyanosis over the past two hours. She had a lifelong history of recurrent cyanosis, a habit of squatting during episodes of breathlessness, and limited physical activity but had never been diagnosed with congenital heart disease. Electrocardiography revealed right axis deviation and right ventricular hypertrophy, while a chest X-ray showed cardiomegaly with a boot-shaped heart silhouette. A hypoxia test supported the suspicion that her hypoxia was cardiac in origin. Initial management included the knee-chest position, high-flow oxygen therapy, intravenous fluids, morphine, and norepinephrine to increase systemic vascular resistance. Subsequent echocardiography confirmed pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA + VSD + MAPCAs), a rare variant of tetralogy of Fallot. **Conclusion**: Hypoxic spells in adults are extremely rare and often underrecognized, especially in resource-limited settings. Rapid diagnosis and early intervention are crucial to improving outcomes. Referral to a specialized cardiac center is essential for comprehensive evaluation and definitive treatment planning.

Keywords: Congenital heart disease, hypoxia, hypoxic spells.

ABSTRAK

Pendahuluan: Spel hipoksik merupakan komplikasi serius dan berpotensi fatal dari penyakit jantung bawaan sianotik. Kondisi ini umumnya terjadi pada anak-anak dan sangat jarang ditemukan pada orang dewasa. Spel hipoksik disebabkan oleh penurunan mendadak aliran darah ke paru, yang menyebabkan hipoksia berat, sianosis, kejang, bahkan kematian. Meskipun jarang, spel hipoksik tetap perlu dipertimbangkan sebagai diagnosis banding pada pasien dewasa yang mengalami krisis hipersianotik, terutama pada mereka yang belum diketahui memiliki riwayat penyakit jantung bawaan. **Kasus**: Seorang wanita berusia 20 tahun datang dengan keluhan sesak napas dan peningkatan sianosis dalam 2 jam terakhir. Ia memiliki riwayat sianosis berulang sejak kecil, kebiasaan jongkok saat mengalami sesak napas, serta keterbatasan dalam aktivitas fisik, namun belum pernah didiagnosis menderita penyakit jantung bawaan. Hasil elektrokardiografi menunjukkan deviasi aksis ke kanan dan hipertrofi ventrikel kanan, sedangkan foto toraks menunjukkan kardiomegali dengan siluet jantung berbentuk sepatu bot. Uji hipoksia mendukung dugaan bahwa hipoksianya berasal dari kelainan jantung. Penanganan awal meliputi posisi lutut-dada, terapi oksigen aliran tinggi, pemberian cairan intravena, morphine, dan norepinephrine untuk meningkatkan resistensi vaskular sistemik. Pemeriksaan ekokardiografi selanjutnya mengonfirmasi adanya atresia pulmonal dengan defek septum ventrikel dan kolateral aortopulmonal mayor (PA + VSD + MAPCAs), suatu varian langka dari tetralogi Fallot. **Kesimpulan**: Spel hipoksik pada orang dewasa sangat jarang dan sering kali tidak dikenali, terutama di fasilitas dengan keterbatasan sumber daya. Diagnosis yang cepat dan intervensi dini sangat penting untuk meningkatkan luaran pasien. Rujukan ke pusat layanan jantung spesialis sangat diperlukan untuk evaluasi menyeluruh dan perencanaan terapi definitif. **Angga Putra Perdana. Spel Hipoksik pada Dewasa: Laporan Kasus.**

Kata Kunci: Penyakit jantung bawaan, hipoksia, spel hipoksik.

Introduction

A hypoxic spell is characterized by a sudden decrease in blood flow to the lungs, which can occur due to an infundibular spasm or a change in the ratio of systemic vascular resistance (SVR) to pulmonary vascular resistance (PVR).[1] This condition can lead to symptoms such as shortness of breath, hypoxia, increased cyanosis, anxiety, seizures, and even death.[2]

Several types of cyanotic congenital heart disease can cause hypoxic spells, including tetralogy of Fallot (TOF), ventricular septal defect (VSD) with pulmonary stenosis (PS) or pulmonary atresia (PA), double outlet right

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ventricle (DORV) with PS and VSD, tricuspid atresia (TA) with VSD and PS, and transposition of the great arteries (TGA).[3] Tetralogy of Fallot is the most common cyanotic heart disease associated with hypoxic spells, but rarely persists into adulthood. Most patients succumb during childhood due to heart failure, severe hypoxia, or related complications. However, in milder cases with better pulmonary blood flow, some individuals can survive into adulthood. The life expectancy for these patients is 49% at 3 years of age, 24% at 10 years, and less than 2% for those over 40 years old.[4,5] Despite the rarity in adults, hypoxic spells should still be considered as a differential diagnosis for hypercyanotic crises to ensure proper management.[5-7]

Case

Our hospital received a referral from a primary health facility for a 20-year-old woman with shortness of breath and cyanosis around her lips and nails. The patient's cyanosis has worsened over the past two hours, preceded by coughing and fever four days ago. She also experienced vomiting eight times today. The patient has a history of recurring cyanosis since birth, squatting habit during dyspnea and has limited physical activities. She has never been diagnosed with congenital heart disease, as she has never undergone further examination at a hospital. Her parents believed that the recurrent cyanosis was normal. There is no family history of similar conditions, and her mother maintained regular health check-ups during pregnancy, with no reported complications.

At the initial presentation, the patient appeared thin and apathetic, with a body weight of 40 kg. Her blood pressure was 68/45 mmHg, with an irregular heart rate ranging from 54 to 59 bpm, a respiratory rate of 26 breaths per minute, a body temperature of 38.1°C, and an oxygen saturation of 59% on room air, increased to 64% with a non-rebreathing mask (NRM) at 15 L/min. Additional findings included slightly elevated jugular venous pressure, rough rhonchi on auscultation in both lung fields, and a fine heart murmur at the upper left sternal border. The patient also had cold extremities and clubbing fingers **(Figure 1).**

Electrocardiography revealed sinus bradycardia with a heart rate of 56 bpm, right axis deviation (RAD), and right ventricular hypertrophy (RVH) **(Figure 2).** A chest x-ray showed cardiomegaly, a boot-shaped heart, and signs of pneumonia **(Figure 3).** Blood tests indicated leukocytosis, metabolic acidosis, and hypoxia **(Table).**

Diagnosis of a hypoxic spell in this patient was based on the patient's complaints of severe hypoxia, a history of cyanosis since birth, a squatting habit during episodes of dyspnea, clubbing fingers, and murmurs on auscultation. ECG and chest x-ray findings supported our assumption of association with cyanotic congenital heart disease. Tetralogy of Fallot (TOF) was suspected, with differential diagnoses include pulmonary atresia with ventricular septal defect (PA+VSD) and concurrent lung infection.

Treatment focuses on reducing oxygen consumption, enhancing oxygen binding, and decreasing right-to-left shunt flow by lowering pulmonary vascular resistance and increasing systemic vascular resistance (SVR). To elevate SVR, the patient was initially placed in the knee-chest position, followed by oxygen 15 L/min via a non-rebreathing mask (NRM).[3,8] An initial of 1500 ml bolus of Ringer's Lactate (RL) was administered, but did not significantly raise the blood pressure.

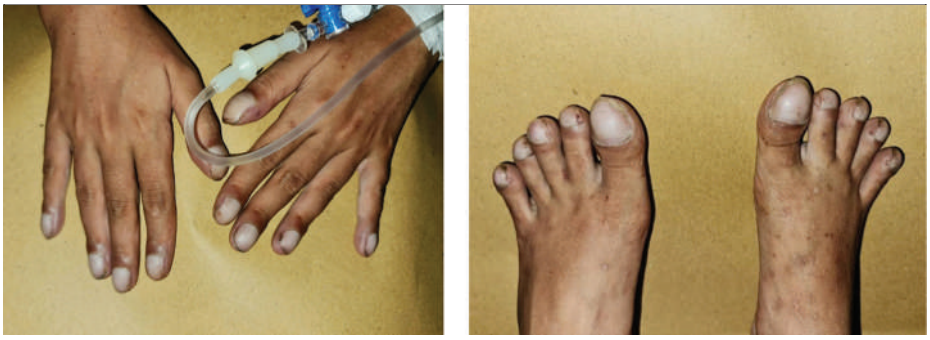


Figure 1. Clubbing fingers.

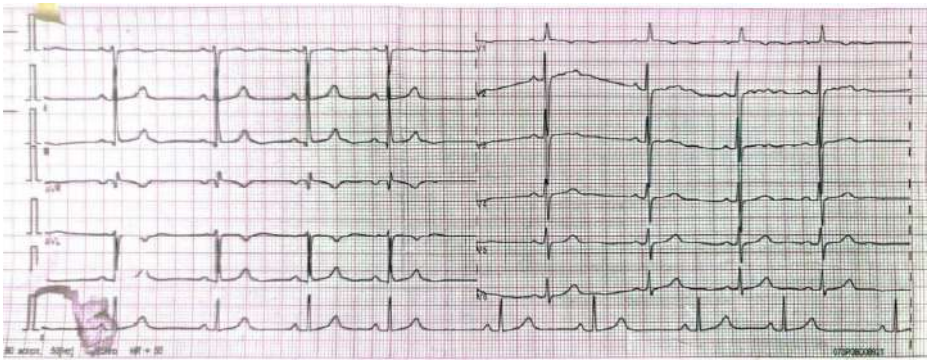


Figure 2. Electrocardiography in the emergency room.

Table. Blood test results.

Complete Blood Count		Blood Chemistry Test		Arterial Blood Gas (ABG)	
Hemoglobin	13.8 g/dL	Blood glucose	102 g/dL	Lactic acid	0.80 mg/dL
White blood	14,350/mm^3	Sodium	142 mmol/L	pH	7.336
Platelets	286,000/mm^3	Potasium	4.3 mmol/L	pCO_2	48 mmHg
Hematocrit	43/mm^3	Chloride	108 mmol/L	BE	-7
	0%	Ureum	35 mg/dL	HCO_3	18.6
	0%	Creatinine	0.8 mg/dL	TCO_2	20 mmol/L
Differential count	2%			SO_2	62%
	89%				
	7%				
	2%				

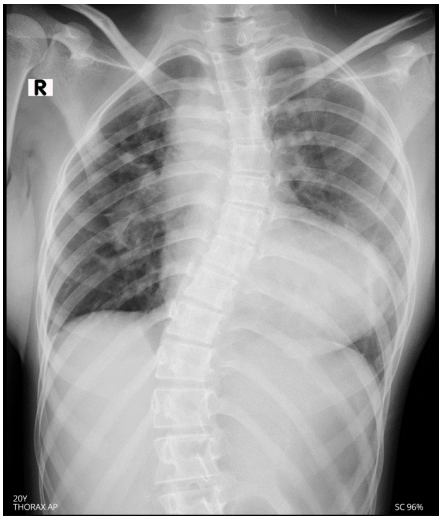


Figure 3. Chest x-ray shows cardiomegaly with a boot-shaped heart, pneumonia, and scoliosis thoracalis.

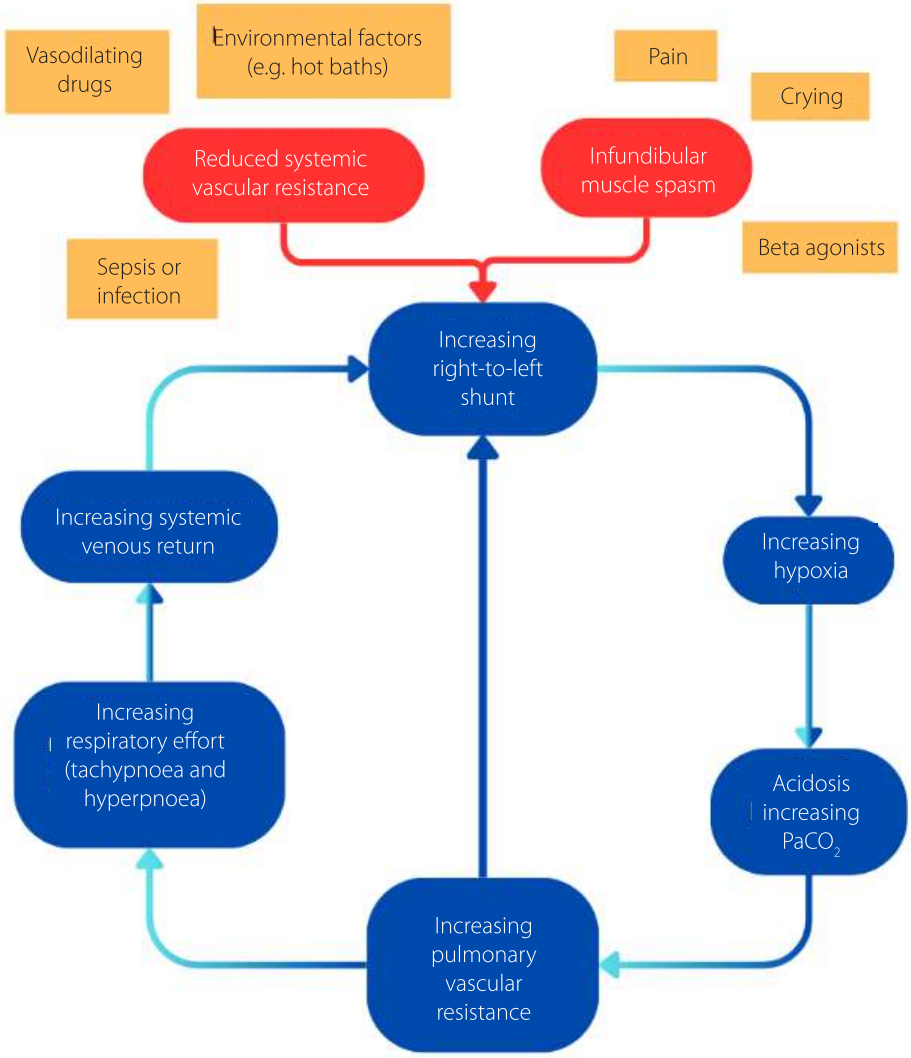
Subsequently, 5 mg of intravenous morphine was given for sedation and to assist in reducing pulmonary vascular resistance (PVR).[3,8] Intravenous norepinephrine was then started at a dose of 0.2 mcg/kg/min (within a range of 0.05 mcg/kg/min to 1 mcg/kg/min) as a vasopressor to increase SVR.[3,8] The patient's condition gradually improved, as evidenced by oxygen saturation increase to 79–88% and blood pressure stabilization at 103/60 mmHg. The patient was then admitted to the ICU for further monitoring. Additionally, we educated the patient's family for referral to Dr. M. Djamil Padang Hospital for more comprehensive evaluation and treatment. Echocardiograph at Dr. M. Djamil Padang Hospital confirmed that a pulmonary atresia, a ventricular septal defect, and major aortopulmonary collateral arteries (PA, VSD, and MAPCAs).

Discussion

The primary diagnosis was hypoxic spells associated with cyanotic congenital heart disease, suspected to be tetralogy of Fallot (TOF), with alternative diagnoses including pulmonary atresia with ventricular septal defect (PA+VSD) and lung infections. Diagnosis was based on severe hypoxia, a history of cyanotic episodes since birth, squatting habit during dyspnea, limited physical activity, clubbing finger, and murmurs in auscultation. Lack of clinical improvement despite oxygen administration supports the suspicion of severe hypoxia related to cardiac issues.[7] The ECG and chest x-ray findings, which reveal an enlarged right heart and a boot-shaped heart silhouette, further confirm the diagnosis.

Diagnosis of hypoxic spells involves a thorough anamnesis, physical examination, and analysis of blood gases. It is important to investigate the patient's history of congenital heart disease with cyanotic episodes and any patterns of squatting following physical activity. Additionally, factors that may trigger hypoxic spells, such as prolonged crying in children, strenuous physical activity, fever, anemia, infection, dehydration, hypoglycemia, and metabolic acidosis (**Scheme 1**)[1] should be explored.[7]

Hypoxic spells are life-threatening, and further examination should not delay the initial therapy. The primary goals are to reduce oxygen consumption, enhance oxygen binding, and decrease right-to-left shunt flow by reducing systemic venous return and increasing blood flow to the lungs.[1,8] The first intervention involves administering 100% oxygen through a non-rebreathing mask (NRM) at 15 L/min and placing the patient in a knee-to-chest position to compress the femoral arteries. This maneuver increases systemic vascular resistance and reduces the right-to-left shunt.[3,8,9] Crystalloid fluids (10–30 ml/kg body weight) are administered to increase preload, which helps enlarge the right ventricular outflow tract (RVOT) and improve blood flow to the lungs. Morphine is also given to alleviate pain and anxiety, reduce heart rate, and induce mild hyperventilation. Morphine helps decrease venous return, reduce pulmonary vascular resistance, and minimize catecholamine release, which can



Scheme 1. Mechanism of spell hypoxia.[1]

mitigate infundibular spasm.[1,3,8,9]

Vasopressors are utilized to increase systemic vascular resistance (SVR). In this case, norepinephrine was chosen due to limited resources. According to the clinical practice guidelines of the Indonesian Association of Cardiologists, vasopressors for hypoxic spells may include phenylephrine infusion at 2-5 µg/kg body weight per minute or intramuscularly at 0.1 mg/kg body weight. Alternatively, metaraminol can be administered as a bolus at 50 mg/100 mL. Norepinephrine infusion at a dose of 0.05-1 µg/kg per minute is also an option, with careful monitoring of blood pressure and patient symptoms. In this case, norepinephrine administration yielded positive results, as evidenced by improvements in blood pressure and oxygen saturation.[3,8]

If severe and persistent hypoxic spells lead to metabolic acidosis, slow intravenous administration of sodium bicarbonate (3-5 mEq/kg body weight) can correct the acidosis. Propranolol (0.1 mg/kg body weight) or esmolol (0.5 mg/kg body weight), followed by a continuous infusion of 50–300 µg/kg/min, may be considered.[3,8] In cases of recurrent spells, propranolol can help reduce infundibular spasm by suppressing contractility and slowing the heart rate, which allows for increased preload and a larger right ventricular outflow tract (RVOT) diameter.[8,10] Sodium bicarbonate and propranolol were not administered in our case due to the mild acidosis and bradycardia, as propranolol could potentially worsen the condition. In cases that do not respond to medical therapy and are accompanied by respiratory failure, immediate mechanical ventilation and surgical intervention may be required.[3,8,9]

Hypoxic spells occur when there is a decrease in systemic vascular resistance accompanied by an increase in pulmonary vascular resistance. In this case, a lung infection may have triggered the hypoxic spells, as suggested by leukocytosis and evidence of pneumonia on the chest x-ray.[1] To address the underlying infection, the patient was treated with ceftriaxone 2 grams once daily, levofloxacin 750 mg once daily, bromhexine 4 mg three times daily, paracetamol 500 mg three times daily, omeprazole 40 mg once daily, and ondansetron 4 mg twice daily. The patient and their family were informed of the need for further comprehensive evaluations and treatments at more advanced facilities.

Hemodynamic disorders resulting from heart defects can manifest with various symptoms, including growth disorders, recurrent cyanosis, limited physical activity, frequent respiratory infections, and heart murmurs—all of which can be early indicators of congenital heart defects.[7,11] If cyanosis is detected, it is essential to perform a blood gas analysis and hypoxic test. Hypoxic test can be conducted

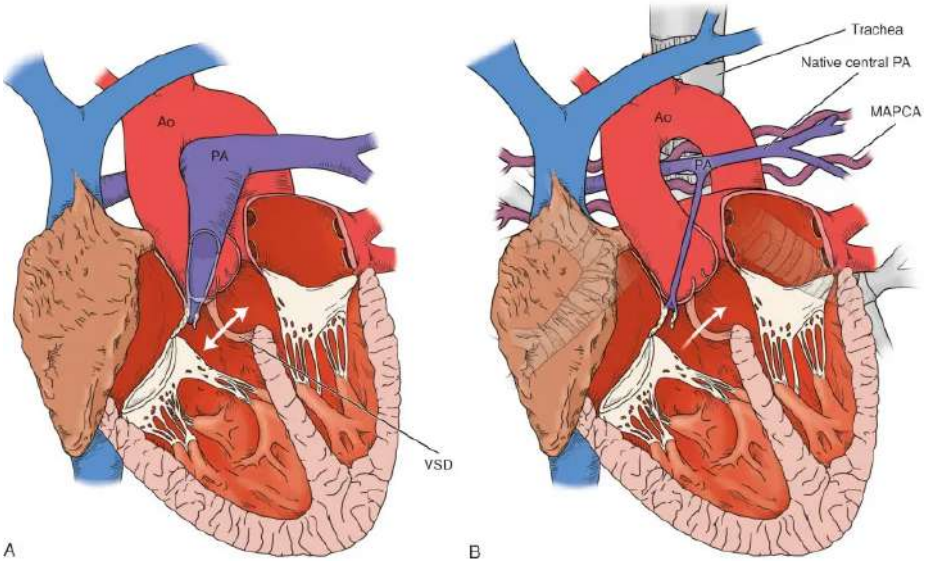
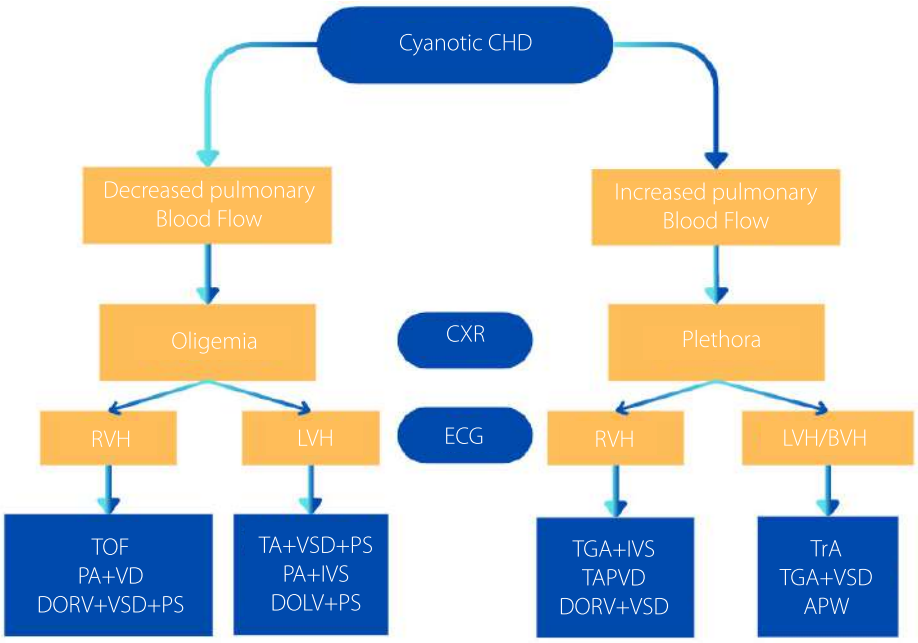


Figure 4. Anatomical features: **A.** Tetralogy of Fallot (TOF), **B.** Pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral artery (PA+VSD+MAPCAS).[12]



Scheme 2. Differential diagnosis of congenital heart disease using chest x-ray and ECG.[13]

Abbreviation: APW= Aortopulmonary window, BPH= Biventricular hypertrophy, CHD= Congenital heart disease, CXR= Chest x ray, DOLV= Double-outlet left ventricle, DORV= Double-outlet right ventricle, ECG= Electrocardiography, IVS= Intact ventricular septum, LVH= Left ventricular hypertrophy, PS= Pulmonary stenosis, PA= Pulmonary atresia, RVH= Right ventricular hypertrophy, TA= Tricuspid atresia, TAPVD= Total anomalous pulmonary venous drainage, TGA= Transposition of great arteries, TOF= Tetralogy of Fallot, TrA= Truncus arteriosus, VSD= Ventricular septal defect.


by administering hyperventilation with 100% FiO_2 oxygen for 10 minutes to observe the increase in PaO_2 through blood gas analysis. A minimal elevation in PaO_2, with a value of PaO_2 <70 mmHg or an increase of <30 mmHg compared to the pre-oxygenation PaO_2 test, strengthens the suspicion of a heart defect.[7] The hypoxia test is particularly valuable for hospitals without access to echocardiography. An ECG and chest x-ray can further aid in confirming the diagnosis by showing enlargement of the right and/or left sides of the heart, depending on the underlying cause.[7,11]

Electrocardiography revealed right axis deviation (RAD) and right ventricular hypertrophy (RVH), indicating enlargement of the right heart, which was confirmed by chest x-ray findings of cardiomegaly, characterized by a boot-shaped heart and an elevated heart apex. These findings suggest cyanotic congenital heart disease, specifically tetralogy of Fallot (**Scheme 2**).[1,7,13] Other differential diagnoses include pulmonary atresia and ventricular septal defect.[1,7] An echocardiogram is necessary to confirm the .defect as shown in their anatomical features (**Figure 4**).[1,7,12] The enlargement of the right heart may result from increased right ventricular pressure, required to maintain pulmonary blood flow. This condition leads to changes in the size of the right ventricular cavity and its muscle mass.[1]

Echocardiography at RSUP M Djamil Padang revealed pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries (PA+VSD+MAPCAs). This condition shares a similar mechanism with tetralogy of Fallot (TOF) but with some anatomical variations, and may produce similar results on ECG and chest x-ray, such as right heart enlargement and a boot-shaped cardiac silhouette. Some experts refer to this condition as tetralogy of Fallot with pulmonary atresia (PA) and major aortopulmonary collateral arteries (MAPCAs).[9,12,14] PA+VSD+MAPCAs is a rare form of cyanotic congenital heart disease. Pulmonary circulation in these patients depends on the extent of pulmonary atresia and the development of collateral vessels. Patients with MAPCAs often show no symptoms at birth because the collateral arteries provide sufficient pulmonary blood flow.[15] For further management, a detailed evaluation of patients with PA+VSD+MAPCAs is essential in determining the appropriate surgical approach. Advanced imaging, such as a CT angiogram or catheter angiography, is recommended for follow-up examinations.[12,16]

Conclusion

Hypoxic spells are life-threatening conditions; early recognition and proper therapy are crucial, especially in the emergency room. Limited time for a thorough examination and inadequate facilities pose additional challenges in diagnosis, particularly in cases without a prior diagnosis of congenital heart disease. Despite their rarity, hypoxic spells should still be considered as a differential diagnosis in cases of hypercyanotic crises.

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