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Case Report

A 13 Year-old Girl with Complete AVSD (Intermediate subtype) and Total AV Block

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ABSTRACT

Atrioventricular septal defects (AVSD) constitute a spectrum of anomalies caused by abnormal endocardial cushion defects. Many classifications have been used to describe AVSD. There is generally subclassified into complete and partial forms. Complete AVSD is characterized by a primum atrial septal defect (ASD) contiguous with an inlet ventricular septal defect (VSD) and a common AV valve. Intermediate AVSD is a subtype of complete AVSD that has distinct right and left atrioventricular valve orifices by a bridging tongue despite having only one common annulus. In some children or adults, we may see unoperated partial or intermediate AVSD, which may be asymptomatic or may present with congestive heart failure, exertional limitation, pulmonary hypertension, infective endocarditis, or heart rhythm disorder. In some cases of AVSD has atrioventricular (AV) conduction disorder, mostly on first-degree AV block. We report a case of a 13-year-old girl with complete AVSD (intermediate subtype) and pulmonary hypertension who is concomitant with total AV block. The patient has undergone implantation of a permanent pacemaker for total AV block and conservative therapy for complete AVSD. Anatomical assessment by trans-thoracal echocardiography (TTE) was essential for diagnostic and detailed morphological characterization of AVSD.

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INTRODUCTION

Atrioventricular septal defect (AVSD) represents a spectrum of congenital anomalies stemming from abnormal development of the endocardial cushion. This condition is typically categorized into two primary types: partial and complete. The complete form of AVSD is characterized by the presence of a primum atrial septal defect (ASD) contiguous with an inlet ventricular septal defect (VSD), absence of fusion in the central leaflet, and a singular orifice comprising the common atrioventricular (AV) valve. Intermediate AVSD, a subtype of complete AVSD, is distinguished by a singular AV valve annulus featuring a "bridging tongue" of tissue that partitions the common AV valve into two distinct orifices (Poterucha et al., 2015; Cetta et al., 2008).

The incidence of AVSD is approximately 4% among all congenital heart diseases (CHD)—notably, 35% of patients with AVSD present with Down syndrome. AVSD may co-occur with Tetralogy of Fallot and other congenital cardiac anomalies. The clinical course of patients with partial or intermediate AVSD who do not undergo surgical intervention can range from asymptomatic to symptomatic, presenting with congestive heart failure, exercise intolerance, pulmonary arterial hypertension (PAH), cyanosis, infective endocarditis, or arrhythmias (Cetta et al., 2008). Echocardiography is a crucial imaging modality in diagnosing AVSD, elucidating the characteristics and morphology of the defect, and assessing its severity (Poterucha et al., 2015).

CASE REPORT

Patient ZA, a 13-year-old male weighing 20 kg, presented to the emergency department of Dr. Soetomo General Hospital in Surabaya with complaints of dyspnea, weakness, and chest discomfort, persisting for one week before admission, worsening one day before admission. The patient reported frequent episodes of dyspnea upon exertion over the past year. He has a history of congenital heart disease diagnosed at the age of one, but he has not been under regular medical supervision, nor has he taken any medication. There is no history of cyanosis or dyspnea at birth. The patient frequently experienced cough, runny nose, and fever at the age of one. The patient and family have approved this case report with informed consent.

Physical examination showed a generally weak condition without signs of anemia or jaundice, with mild cyanosis observed. Vital signs included a blood pressure of 90/60 mmHg, a heart rate of 52 beats per minute (regular), a respiratory rate of 20 breaths per minute, an axillary temperature of 36.8°C, and an oxygen saturation of 90%. Cardiopulmonary examination showed an ictus cordis at the fifth intercostal space (ICS) 1 cm lateral to the left mid-clavicular line, single S1 and S2 heart sounds, and a grade III/VI systolic murmur at the fourth ICS along the left parasternal line. Breath sounds were vesicular without rhonchi or wheezing. The extremities were warm, with no edema.

Laboratory results suggested hemoglobin (Hb) of 16.8 g/dL, hematocrit (Ht) of 49%, white blood cell count (WBC) of 10,800/uL, platelets of 123,000/uL, sodium of 134 mEq/L, potassium of 3.7 mEq/L, chloride of 104 mEq/L, blood urea nitrogen (BUN) of 11 mg/dL, serum creatinine (SC) of 0.48 mg/dL, aspartate aminotransferase (SGOT) of 49 u/L, and alanine aminotransferase (SGPT) of 25 u/L.

Initial electrocardiography showed a sinus rhythm at 70 bpm, total atrioventricular block, junctional escape rhythm at 52 bpm, and non-

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specific ST depression in leads V2-V5 (Figure 1.1). A chest radiograph revealed cardiomegaly with a cardiothoracic ratio (CTR) of 79%, a prominent pulmonary conus, and increased pulmonary vascularity (Figure 2)

Subsequently, Temporary Pacemaker (TPM) placement was performed on August 2, 2016, due to the indication of total AV block, with TPM settings at HR 80 bpm, Output 3 mA, and Sensitivity 3 mV. Post-TPM placement EKG can be seen in Figure 1.2.

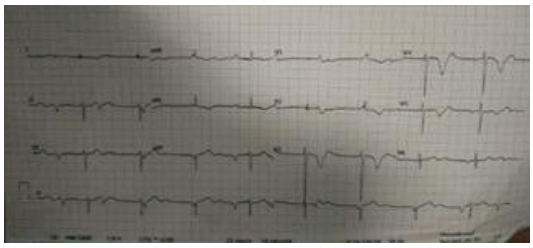

Figure 1.1 Initial ECG

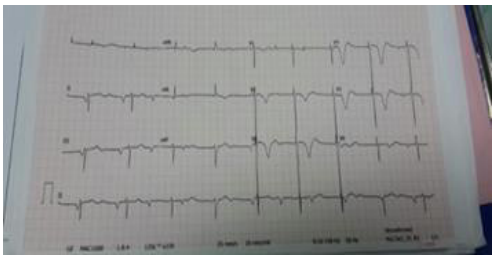

Figure 1.2 ECG post TPM placement

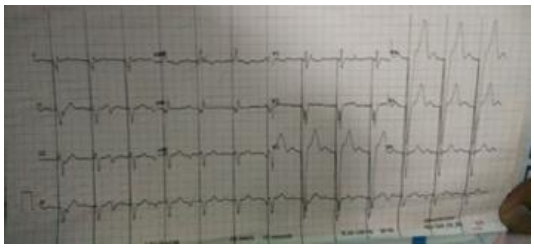

Figure 1.3 ECG 8 Weeks After TPM Placement

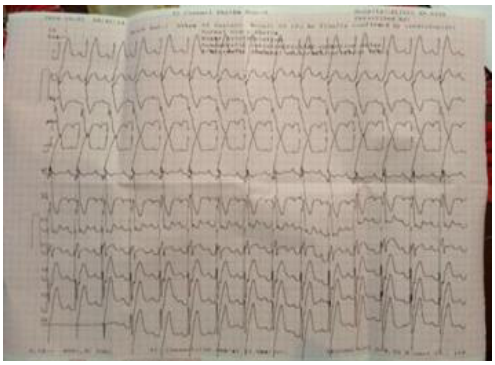

Figure 1.4. ECG post PPM Placement

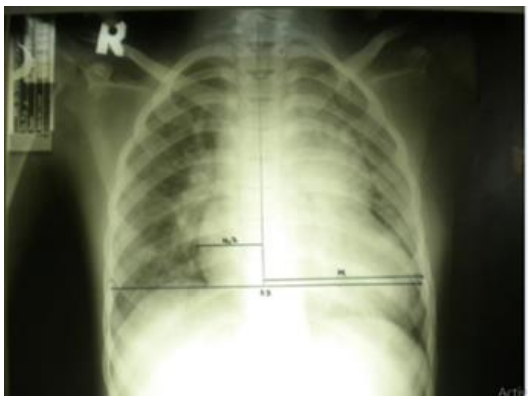

Figure 2. Thorax Photo Imaging (X-Ray)

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From the trans-thoracic echocardiography (TTE) examination, the findings revealed an ambiguous site, AV-VA concordance, normal pulmonary vein drainage, dilatation of all cardiac chambers, severe mitral (left AV valve) regurgitation (maxPG 83.29 mmHg), trivial tricuspid (right AV valve) regurgitation, a large primum ASD with a diameter of 5.18 cm, functional single atrium, a large inlet VSD with a diameter of 1.01 cm, normal LV systolic function (LVEF 63.84%), and a left-sided aortic arch. The conclusion is Complete AVSD-Intermediate type (see figures 3.1-3.4)

The patient was subsequently admitted for condition improvement and observation, with initial therapy consisting of diuretics (furosemide), inotropes (dobutamine), and TPM support. On August 30, 2016, right and left heart catheterization (DxRL) was performed, revealing an ambiguous site (left isomerism), Absence of innominate vein, Persistent LSVC visible towards RA (bilateral SVC), VSD with a diameter of 1.5 cm ± 0.9 mm, MR grade II-III visible, and a goose neck appearance. The conclusion is Ambiguous site (left isomerism) + absence of innominate vein + visible Persistent LSVC + AV canal defect with MR grade II-III (see figure 4).

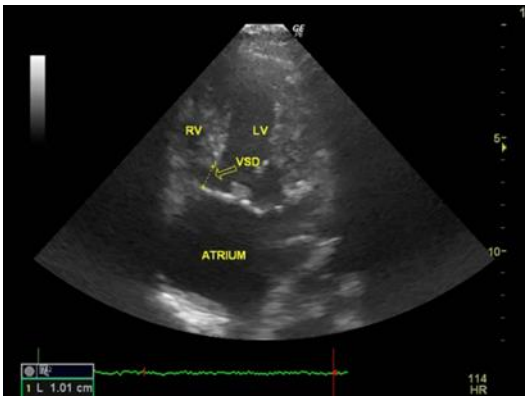

Figure 3.1 Echo A4C: Large Inlet VSD

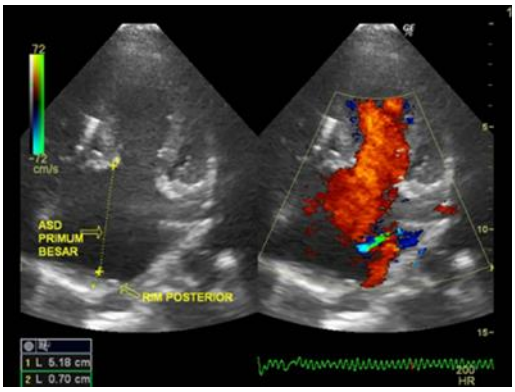

Figure 3.2 Echo A4C: Large Primum ASD

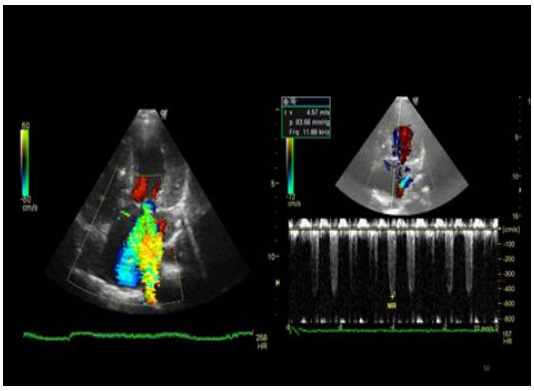

Figure 3.3 Mitral/ left AV Valve Rergitation

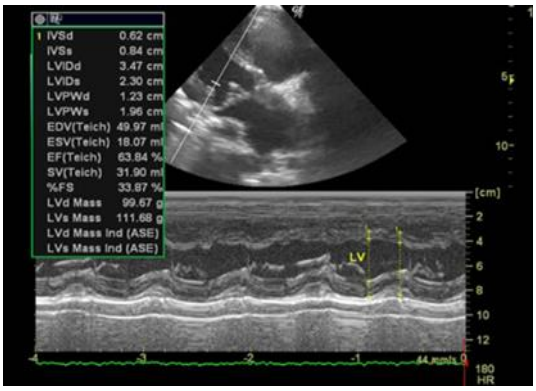

Figure 3.4. LV Study: EF 63,84%

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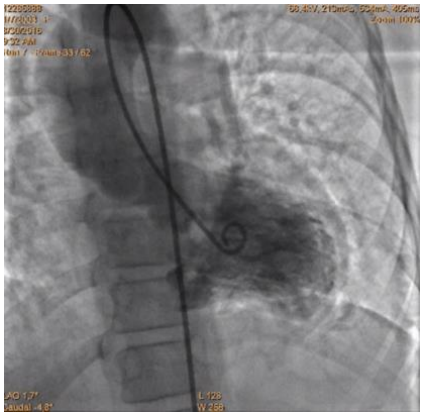

Figure 4. Left Ventriculography showed a gooseneck appearance

Based on the anamnesis, physical examination, and supporting investigations, a diagnosis of Complete AVSD - intermediate type accompanied by severe PH and Total AV block was established. After more than 8 weeks of observation following TPM implantation, the patient's TAVB rhythm on the EKG remained unchanged (see Figure 1.3). Through a discussion at the integrated heart care center, it was decided to proceed with the intravenous implantation of a Permanent Pacemaker (PPM) and conservative therapy for their Complete AVSD.

The PPM was implanted on September 27, 2016, using a St. Jude Microny II pacemaker with VVIR lower rate set at 80 bpm, amplitude 2.1 mA, sensitivity 4.0 mV, intravenously via the right subclavian vein. (See post-PPM EKG results in Figure 1.4). The patient was discharged in stable condition on October 2, 2016, and received therapy including diuretics, PDE-5 inhibitor, and antibiotics.

DISCUSSION

Atrioventricular Septal Defect (AVSD)

Terminology and Classification

Atrioventricular septal defect (AVSD) is part of the spectrum of anomalies caused by abnormal development of the endocardial cushion defect. AVSD is also known as 'AV canal defect', 'persistent common AV canal', or 'endocardial cushion defect'. There are many classifications of AVSD, making it challenging to summarize AVSD terminology. In general, it is classified into two types: complete and partial. Complete AVSD is characterized by a primum ASD adjacent to the VSD inlet, no central fusion of valve leaflets with only one orifice present, and a common AV valve.

In contrast, partial AVSD differs from complete AVSD because the malformation does not include a VSD inlet. The anatomical description of partial AVSD includes a primum ASD and an anterior cleft in the mitral valve leaflet. In this type, the mitral and tricuspid valves each have complete and separate annuli (Poterucha et al, 2015).

Epidemiology

AVSD represents 4-5% of all congenital heart defects, with a prevalence ranging from 0.19 per 1000 live births. It occurs with equal distribution between genders or slightly more in females. About 40-45% of children with Down syndrome have congenital heart defects, and about 40% of them have AVSD, usually the complete type. Complete AVSD is also commonly found in patients with heterotaxy syndrome (Olariu, I. C., et.al, 2021).

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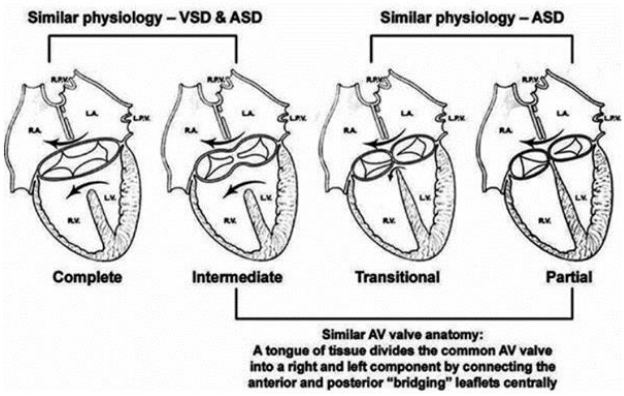

Figure 5. AVSD Classification Summary (Poterucha *et al*, 2015)

Clinical Manifestations

The course of AVSD disease in reaching adulthood, whether partial or intermediate type without surgery, can be asymptomatic or symptomatic. Clinical symptoms of AVSD are non-specific due to intracardiac shunts (L-R, R-L, or bidirectional), which may manifest as congestive heart failure, activity intolerance, pulmonary arterial hypertension (PAH), cyanosis, infective endocarditis, or cardiac arrhythmias (atrial fibrillation/atrial flutter). In some cases, AVSD disease may not extend to adulthood. The clinical presentation depends on the presence and size of ASD and VSD, and the competence of the left AV valve (Shimbori, R., Takaki, J., et.al, 2021)

Physical Examination

In patients with complete AVSD, hyperactive cardiac impulse can be found, and the systolic murmur is loudest at the lower left sternal border, with a third heart sound and an accentuated pulmonary component of S-2 (Spector LG, Menk JS, Knight JH, et al; 2018). In infants, signs of congestive heart failure such as tachycardia, tachypnea, hepatomegaly, and gallop rhythm can be found (Park MK, 2014).

Electrocardiography

Most patients with complete AVSD have a prolonged PR interval (first-degree AV block). A superior axis is often found, with QRS axis between -40 and -150°, sometimes accompanied by right ventricular hypertrophy (RVH) or right bundle branch block (RBBB), and biventricular hypertrophy may also be present (Park MK, 2014).

Chest X-ray

Chest X-ray may reveal cardiomegaly due to enlargement of the right or left heart chambers, depending on the degree and direction of the left-to-right shunt. Pulmonary vascular markings are usually increased, with prominent pulmonary artery segments (Park MK, 2014).

The patient is a 13-year-old female, currently in junior high school, presenting with complaints of easy fatigue during strenuous activities over the past year, without a history of cyanosis since birth. Upon examination, a cardiac impulse is noted at the 5th intercostal space, 1 cm lateral to the mid-clavicular line on the left side. There is a single S1-S2 heart sound and a systolic murmur of grade III/VI at the 4th intercostal space parasternal line on the left side. Electrocardiography reveals a sinus rhythm with TAVB (Third-degree atrioventricular block) and a junctional escape rhythm. Chest X-ray

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findings include cardiomegaly with a prominent pulmonary cone and increased lung vascularity. Based on these findings, there is suspicion of a ventricular septal defect (VSD), which needs to be confirmed via echocardiography.

Trans-Thoracic Echocardiography

TTE holds significant clinical utility and serves as the primary imaging modality for diagnosing AVSD. Broadly, TTE examinations should encompass the evaluation of ASD primum, VSD inlet, morphology and function of the AV valves, cardiac chamber dimensions, dominant shunt direction (L to R; R to L, bidirectional), and subaortic stenosis (if present).

Evaluation of echocardiographic parameters in AVSD (Poterucha et al, 2015):

- Assessment of ASD primum, visualized in subcostal four-chamber (4C), parasternal, and apical 4C projections. In the A4C projection, ASD primum and the insertion of the tricuspid and mitral valves at the apex of the septum can be observed.
- Assessment of VSD inlet, visible in the apical 4C and subcostal 4C positions.
- AV valve morphology and the parallel insertion of left-right AV valve components at the same level can be evaluated through apical 4C projection and AV valve attachment.
- Assessment of 'Cleft' anterior mitral leaflet.
- Assessment of the degree of regurgitation of both AV valves.
- Assessment of the presence or absence of subaortic stenosis, which can develop progressively even after surgery.
- Evaluation of LVOT elongation, indicating a longer LV outlet than inlet in the Plax view, where LVOT is narrow and long or shows a "goose neck" appearance.
- Evaluation of the 'balance' between the right and left heart chambers.

In this patient's TTE, we observe the appearance of a common atrioventricular valve with two orifices (right and left AV valves) with tissue separating the right and left AV valves. There is severe mitral (left AV valve) regurgitation (maxPG 83.29 mmHg) and suspicion of a cleft anterior mitral leaflet, trivial tricuspid (right AV valve) regurgitation, a large ASD primum with a diameter of 5.18 cm (functional single atrium), a large VSD inlet with a diameter of 1.01 cm, elongation of the LVOT, and a "goose neck appearance." Subaortic stenosis is not observed. Other findings include: ambiguous situs, AV-VA concordance, normal pulmonary vein drainage, dilated chambers in all heart chambers, normal LV systolic function (LVEF 63.84%), and a left-sided aortic arch. Based on the echocardiography results, our patient is classified as having complete AVSD, intermediate subtype.

Cardiac Catheterization

Cardiac catheterization is rarely required for managing infants with complete AVSD, but in older children, it is necessary to assess pulmonary vascular resistance. Cardiac catheterization also allows for the assessment of oxygen saturation and pressures in the cardiac chambers, thus evaluating hemodynamic consequences in patients. Although LV angiography is seldom necessary, it can demonstrate the "gooseneck" appearance and determine the degree of AV valve regurgitation (Rui Anjos, 2021).

In this patient's cardiac catheterization, Situs Ambiguus (left isomerism) was found, with no innominate vein visible, persistent LSVC seen draining into the RA (bilateral SVC), a VSD with a diameter of 1.5 cm ± 0.9 mm, MR grade

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II-III, and a gooseneck appearance. The pressure in the right ventricle was 85/0 mmHg without stenosis in the pulmonary valve. These findings support a diagnosis of complete AVSD with situs ambiguus (left isomerism) and bilateral SVC with pulmonary hypertension.

Management and Treatment

Management of complete AVSD may involve medical therapy and surgical intervention. In infants with congestive heart failure, anti-congestive medications (vasodilators, diuretics) may be administered. However, the definitive therapy for AVSD is surgical intervention. Surgical intervention for AVSD has significantly reduced morbidity and mortality in recent decades. The goal of surgery for complete AVSD is to close the interatrial and interventricular spaces and restore competent mitral valve function to avoid stenosis and conduction tissue damage. Closure of the mitral valve cleft is usually performed if present, and the tricuspid valve is repaired if necessary (Park MK, 2014).

Prognosis

Generally, AVSD patients who survive have undergone surgery in infancy or childhood. Patients with complete AVSD who have not undergone surgery have a poor prognosis, with symptoms such as Eisenmenger syndrome, unless the VSD size is small. In cases of partial AVSD without surgery, symptoms may occur in adulthood. The clinical presentation of partial AVSD depends on the L to R shunt at the atrial level and/or left AV valve regurgitation ('cleft mitral'), often asymptomatic, but symptoms tend to increase with age (Saxena et. al, 2019).

In this patient, surgical intervention was not performed due to the likelihood of existing pulmonary hypertension (as seen on chest X-ray and cardiac catheterization), thus, conservative management was pursued (only medical therapy administered).

Total Atrioventricular Block (TAVB) In Children

Atrioventricular (AV) block is a cardiac electrical disorder characterized by impaired conduction from the atria to the ventricles for various reasons. There are several types of AV blocks, namely first-degree AV block, second-degree AV blocks, and third-degree/total AV block (Yeo et al, 2011). Some children with AV block initially exhibit asymptomatic symptoms, followed by various manifestations such as palpitations, chest pain, weakness, syncope, activity intolerance, and dizziness, and some may experience congestive heart failure in congenital total AV block. Symptoms depend on the ventricular rate, frequency of premature ventricular beats, and atrioventricular synchrony (Singh, 2012).

In our patient, TAVB was detected on EKG, and clinical manifestations of weakness and activity intolerance were observed.

TAVB in Children: Acquired or Congenital?

The causes of AV block in children can be classified as acquired (acquired) or congenital cases. Acquired AV block can result from various extrinsic and intrinsic conditions. Progressive idiopathic degeneration of the cardiac conduction system can occur in about half of AV block cases. The causes of progressive degeneration may be due to neuromuscular disorders (such as muscular dystrophy), systemic diseases (such as sarcoidosis and cardiac amyloidosis), neoplastic disorders (such as primary cardiac lymphoma and/or post-radiation therapy), after catheter ablation of atrial or AV node pathways, after surgery (such as VSD closure surgery), infections (such as Lyme disease), or drug-related causes (such as digitalis, calcium channel blockers) or toxins (Vogler et al, 2012; Saleh et al, 2014).

Meanwhile, congenital AV block can occur in an isolated disease (standalone), usually

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resulting from intrauterine exposure to maternal antibodies (such as Rho antibodies, La antibodies) or cases related to other congenital diseases (Vogler et al, 2012). Total AV block is a fairly common manifestation in the congenitally corrected type of transposition of the great arteries (Vogler et al, 2012). Mutations in various genes can also occur, such as mutations in the NKX2.5 gene (a member of the NK-2 class), which have been shown to be present in autosomal dominant ASD with progressive AV block occurring in 1-4% of sporadic ASD patients (Sarkozy et al, 2005).

TAVB in AVSD

Patients The occurrence of TAVB can be found in patients with complete AVSD. In patients with AVSD, the AV node shifts inferiorly and posteriorly from its normal position in the Koch triangle due to defects in the AV septum, mostly (50%) resulting in first-degree AV block, as it requires more time to pass through the AV node (Calkoen, 2015).

Abnormalities in TAVB in AVSD may also be associated with left isomerism heterotaxy. In left isomerism heterotaxy, AVSD, bilateral superior vena cava, bilateral morphology of the left atrium, atresia or absence of the sinoatrial node, transverse liver, polysplenia, usually common atrium, interruption of the inferior vena cava with continuation of the azygos, partial pulmonary venous connection, LVOT obstruction, VA concordance can be found (Perloff & Merelli, 2011).

In this patient, congenital TAVB is likely, where during an 8-week observation period, factors causing cardiac rhythm disturbances have been ruled out (no infection, normal electrolytes, and no drugs affecting the AV node). In this patient we found evidence of TAVB in a patient with complete AVSD of the intermediate subtype, where there is also left isomerism ambiguous site, interruption of the inferior vena cava, and bilateral SVC, thus leading to congenital TAVB.

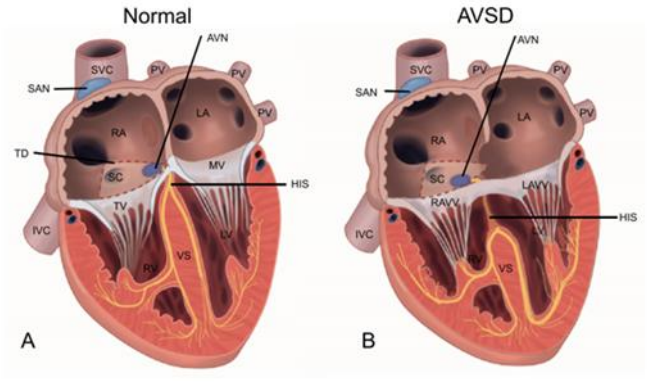

Figure 6. AV node location is more posterior and inferior in AVSD patients compared to normal patients (Calkoen et al, 2015).

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Management of TAVB in Children

TAVB is a serious condition that can disrupt hemodynamics and requires appropriate therapy. Managing children with AV block should begin by identifying potential reversible causes, such as infection, myocardial ischemia, or drugs that interfere with AV node conduction (such as digitalis or beta-blockers). If these drugs are present, they should be discontinued. In cases of acute symptomatic AV block, intravenous vagolytic agents such as atropine and/or catecholamines (adrenaline) can be used. If these treatments are ineffective, temporary pacemaker placement is indicated. Placement of a temporary or permanent pacemaker is the main definitive therapy in most symptomatic TAVB cases (Vogler et. al, 2012).

Permanent Pacemaker Implantation in Children

The goal of pacemaker implantation in children with TAVB is to restore heart rhythm and rate, improve symptoms due to bradycardia, provide hemodynamic stability, and improve quality of life (Baruteau, 2016). The number of children requiring permanent pacemaker implantation is very small compared to adults. Only 1% of pacemaker recipients are children. The implantation of a permanent pacemaker in children requires various considerations and specialized skills of a surgeon, pediatrician, and cardiologist regarding the consequences and prognosis of patients due to lifelong pacemaker use (Silvetti et. al, 2024).

Below are the complete indications for permanent pacemaker implantation in children and adolescents based on the PACES Expert Consensus Statement On The Indications And Management Of Cardiovascular Implantable Electronic Devices In Pediatric Patients in 2021 (Shah et al, 2021):

According to the criteria above, the patient meets the Class I criteria for permanent pacemaker implantation.

	Recommendations		
COR	Atrioventricular Block: Other Considerations	LOE	References
I	Permanent pacemaker implantation is indicated in patients with clinically significant ventricular tachycardia (VT) that is pause dependent or associated with severe bradycardia; ICD implantation may be considered as a reasonable alternative.	C-LD	19
I	Permanent pacing is indicated in *symptomatic* patients with idiopathic advanced second- or third-degree AV block not attributable to reversible causes.	C-LD	
IIa	Permanent pacemaker implantation is reasonable for any degree of AV block that progresses to advanced second- or third-degree with exercise in the absence of reversible causes.	C-LD	20
IIb	Permanent pacemaker implantation may be considered for patients with intermittent advanced second- or third-degree AV block not attributable to reversible causes and associated with minimal symptoms that are otherwise unexplained.	C-LD	
III Harm	Permanent pacemaker implantation is not indicated for asymptomatic first-degree AV block or asymptomatic second-degree Mobitz type I.	C-LD	2,5

Figure 7. PACES Expert Consensus Statement On The Indications And Management Of Cardiovascular Implantable Electronic Devices (Shah et. al, 2021)

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Location of Permanent Pacemaker Generator Implantation in Children

Unlike in adults, permanent pacemaker implantation in children always requires individual assessment to determine the implantation location (infraclavicular, abdominal, or other) and topology (subcutaneous or submuscular) (Antretter et al, 2003).

Selection of Permanent Pacemaker Access: Epicardial or Intravenous?

Several patient conditions and lead factors must be considered when selecting the access route for permanent pacemaker implantation in children (Udink TC & Sreeram, 2011). An epicardial approach is recommended in children up to 3 or 4 years of age to prevent subclavian vein lesions. Meanwhile, an intravenous approach is more advisable in children weighing more than 15 kg, although implantation in smaller patients has been reported (Udink TC & Sreeram, 2011).

Criteria for intravenous pacemaker pacing in children include age ≥ 4 years, weight ≥ 15 kg, absence of right-to-left shunt, adequate superior vena cava flow into the right atrium, and no concurrent heart surgery (Singh HR, 2013). Intravenous pacing offers advantages in preventing thoracotomy or sternotomy complications, better lead longevity, and lower pacing thresholds, thereby preserving battery longevity. However, issues related to intravenous pacing include an increased risk of venous thrombosis and tricuspid valve integrity impairment. Although venous thrombosis is generally asymptomatic, it can manifest as superior vena cava syndrome in some patients. In the future, the discovery of smaller endocardial leads (4.1 F) may reduce the risk of venous thrombosis (venous occlusion) in children undergoing intravenous pacing (Shah et.al, 2021).

The patient was decided to undergo intravenous permanent pacemaker implantation using VVIR mode.

CONCLUSION

This case study reports a 13-year-old female child who has been complaining of easy fatigue during strenuous activities over the past year without a history of cyanosis since birth. Based on history taking, physical examination, and diagnostic tests (ECG, TTE, and cardiac catheterization), we concluded a diagnosis of complete atrioventricular septal defect (intermediate subtype) accompanied by pulmonary hypertension and total atrioventricular block.

Transthoracic echocardiography (TTE) plays a crucial role as the primary imaging modality in confirming the diagnosis. Cardiac catheterization can be useful if there are doubts about the TTE results and to determine pulmonary vascular resistance. Management of complete atrioventricular septal defect (AVSD) may involve medical therapy and surgical intervention. Surgical treatment requires consideration of patient symptoms, pulmonary vascular resistance, and age. Atrioventricular block (AVB) in children can result from acquired and congenital causes. Management of AVB in children may involve medical therapy, and permanent pacemaker implantation is often necessary.

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