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# Late Presentation of Pulmonary Hypertension Crisis Concurrent with Atrial Arrhythmia after Atrial Septal Defect Device Closure

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**Abstract:-**

➢ ***Background:***

**ASD occurs when there is a septal defect between the right and left atria, resulting in a left-to-right shunt that increases the volume of the right heart and pulmonary circulation. Increased pulmonary resistance can lead to pulmonary hypertension (PH), resulting in progressive deterioration of right ventricular function, leading to right heart failure and death. Prolonged elevation of atrial pressure induces progressive atrial dilatation and electrophysiological remodelling. Together with autonomic modulation, this leads to atrial arrhythmias (AAs). Patients with significant shunts leading to ventricular volume overload are considered for ASD closure. However, in some cases, PH occurs after ASD closure.**

➢ ***Case Presentation:***

**We report a 21 yo man diagnosed with ASD Secundum Post Closure with Device (September 1th, 2023) and Pulmonary Hypertension Crisis. The left atria (LA), right atria (RA) and right ventricle (RV) were dilated. We also found moderate mitral regurgitation, severe tricuspid regurgitation, and mild to moderate pulmonary regurgitation. There was a decline in systolic function in the right ventricle, and grade III diastolic dysfunction in left ventricle. There was a well-seated device with no residual shunt on interatrial septal. The pulmonary arteries were confluence and dilated. From ECG we found atrial flutter with variable conduction. This patient was transferred to HCU. This patient treated with digoxin, furosemide, milrinon, ceftriaxone, miniaspi, sildenafil, electrophysiology, and 3D ablation.**

➢ ***Conclusion:***

**Pulmonary hypertension can occur in cases of congenital heart defects, such as ASD. The operative management of ASD is closure of the ASD, but in some unique groups, this can lead to pulmonary hypertensive crisis after its closure.**

***Keywords:-*** *ASD (Atrial Septal Defect); ASD Secundum Post Closure; Pulmonary Hypertension Crisis; Right Heart Failure.*

## I. INTRODUCTION

➢ *Background*

Pulmonary hypertension (PH) is a chronic disorder characterised by elevated resting mean pulmonary arterial pressure (mPAP) ≥ 20 mmHg and can result from several disease processes. Irrespective of its aetiology, PH is a progressive, decompensated disease that is often advanced and has a poor prognosis despite the development of new therapeutic agents

On the other hand, a pulmonary hypertensive crisis is characterized by a sudden and significant rise in the pressure of the pulmonary artery above the mean arterial pressure (MAP). This is produced by a sudden narrowing of the blood vessels in the lungs, leading to the failure of the right ventricle and a decrease in blood pressure throughout the body. Ultimately, this results in a severe lack of oxygen in the tissues and can lead to death.

The incidence of atrial septal defect (ASD) is 1.6 per 1,000 live births, and the proportion of individuals with ASD who survive into adulthood is 97%. Atrial septal defect (ASD) is a condition characterized by a hole in the wall separating the right and left atria of the heart. This hole causes blood to flow from the left atrium to the right atrium, leading to an increase in the volume of the right side of the heart and the circulation of blood in the lungs. Pulmonary hypertension (PH) can occur as a result of elevated pulmonary resistance, leading to a gradual decline in the functioning of the right ventricle. This can ultimately end in right heart failure and mortality. Patients who have large abnormal connections causing excessive blood flow to the ventricles are evaluated for atrial septal defect closure.

In addition, long-term excessive strain on the right ventricle results in the malfunctioning of the tricuspid valve, causing the backward flow of blood and a consequent rise in pressure inside the right atrium. Continued increase in pressure in the atrium causes the atrium to gradually expand and undergo changes in its electrical properties. In conjunction with autonomic regulation, this results in the occurrence of atrial arrhythmias (AAs). The recognition of arrhythmias as a significant factor contributing to illness and death in individuals with pulmonary hypertension has been established for many years. The cohort study found that the yearly occurrence rate of AAs was 2.8% per year, resulting in

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a total occurrence rate of 11.7% during the course of the six-year study. Specifically, there were 12 patients with atrial flutter, 12 with atrial flutter, and three with atrioventricular nodal reentrant tachycardia.

## II. LITERATURE REVIEW

The delayed onset of a pulmonary hypertension crisis concurrent with atrial arrhythmia following atrial septal defect device closure is a significant concern in clinical practice. Pulmonary hypertension can emerge as a severe complication post-closure of atrial septal defects, potentially leading to adverse outcomes such as sudden death (Serinelli et al., 2019). The prevalence of pulmonary hypertension in individuals with open atrial septal defects is substantial, highlighting the critical need for timely intervention and monitoring (Gloan et al., 2018). Left untreated, atrial septal defects can result in pulmonary hypertension in a considerable proportion of patients, particularly in adulthood (Lammers et al., 2005).

Complications like atrial arrhythmias may develop after the closure of atrial septal defects, emphasizing the importance of appropriate management strategies (Varma et al., 2003). Moreover, the presence of a deficient retro-aortic rim is a risk factor for complications such as aortic erosion following device closure of atrial septal defects, underscoring the significance of meticulous procedural planning (O'Byrne et al., 2014).

In certain instances, the closure of atrial septal defects in the presence of severe pulmonary hypertension remains a topic of debate due to the potential risks associated with a sudden increase in pulmonary pressure post-closure (Fu et al., 2023). Additionally, the closure of large defects in elderly patients necessitates thorough evaluation to prevent complications like increased left atrial pressures (Holzer et al., 2005).

Late complications such as bi-atrial thrombus formation, cerebral infarction, and pulmonary embolism can occur following atrial septal defect closure, highlighting the need for vigilant monitoring and management (Xiong et al., 2022). Systemic desaturation and exertional dyspnea may also manifest years after device closure of atrial septal defects, emphasizing the importance of long-term follow-up and surveillance (Bagaria & Hiremath, 2022).

In conclusion, the delayed onset of pulmonary hypertension crises concurrent with atrial arrhythmia after atrial septal defect device closure stresses the importance of comprehensive pre-procedural assessment, meticulous procedural techniques, and long-term monitoring to mitigate potential complications and ensure optimal patient outcomes.

## III. CASE PRESENTATION

A 21-year-old man presented to the PJT with a 1-year history of shortness of breath, especially on exertion. He was admitted with shortness of breath for 2 days, worsening in the last 1 day. He also complains of fatigue and palpitation. The patient had difficulty gaining weight since infancy. The patient was born spontaneously at term, birth weight unknown. The patient was previously admitted to the Cardiac Centre in August 2023 for atrial septal defect closure with subsequent stabilisation and discharge to home for further outpatient management. The patient is currently routinely monitored, taking miniaspi 80mg, bisoprolol 5mg, sildenafil 20mg, furosemide 40mg, lansoprazole 30mg. On presentation, blood pressure was 123/76 mm Hg, pulse 125 bpm, irregular, respiratory rate 26 breaths/min, and saturation different in each limb (right arm 64%, left arm 58%, right foot 62%, and left foot 59%). He had a split-second heart sound and holosystolic murmur at the left lower sternal border (LLSB). Physical examination revealed clubbing fingers without cyanosis of the extremities.

A 12-lead electrocardiogram showed aupraventricular rhythm, heart rate average 120 beats per minute, irregular, right axis deviation (RAD) and right ventricular hypertrophy (RVH) (Figure 1). Chest radiograph showing cardiomegaly (cardiothoracic ratio, 0.79) with a left-to-right shunt consistent with ASD, pulmonary oedema, and an ASD occluder device at CV T7-T9 level (Figure 2). Laboratory findings may reveal leukocytosis and prolonged hemostatic time (Figure 3), elevated liver enzymes, azotemia, electrolyte depletion (Figure 4), and completely compensated respiratory acidosis (Figure 5).

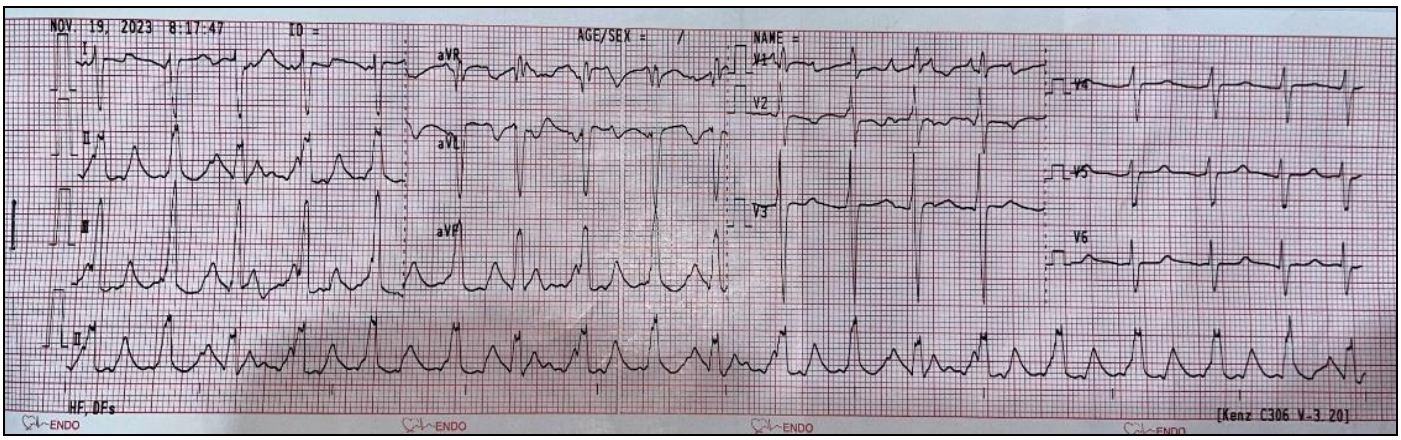


Fig 1 Supraventricular Rhythm, Heart Rate Average 120 Beats per Minute, Irregular, Right Axis deviation (RAD) and Right Ventricular Hypertrophy (RVH)

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Based on Figure 1. This EKG reading points to a potential heart rhythm issue. The source of the electrical signal seems to be above the ventricles, with a faster and irregular heart rate averaging 120bpm. Additionally, right axis deviation and right ventricular hypertrophy suggest the right ventricle may be enlarged, possibly due to increased workload or abnormal electrical conduction. The information describes an abnormal heart rhythm: The electrical signal originates above the ventricles, the lower chambers of the heart. This is in contrast to a ventricular rhythm, where the signal starts in the ventricles; The average heart rate is 120 beats per minute (bpm). A normal resting heart rate is typically between 60-100 bpm; The heart rate is not consistent, meaning the time between beats varies; An ECG (electrocardiogram) reading that suggests the electrical impulse spreads abnormally through the heart, potentially indicating an enlarged right ventricle; The right ventricle, responsible for pumping blood to the lungs, is thickened and enlarged.

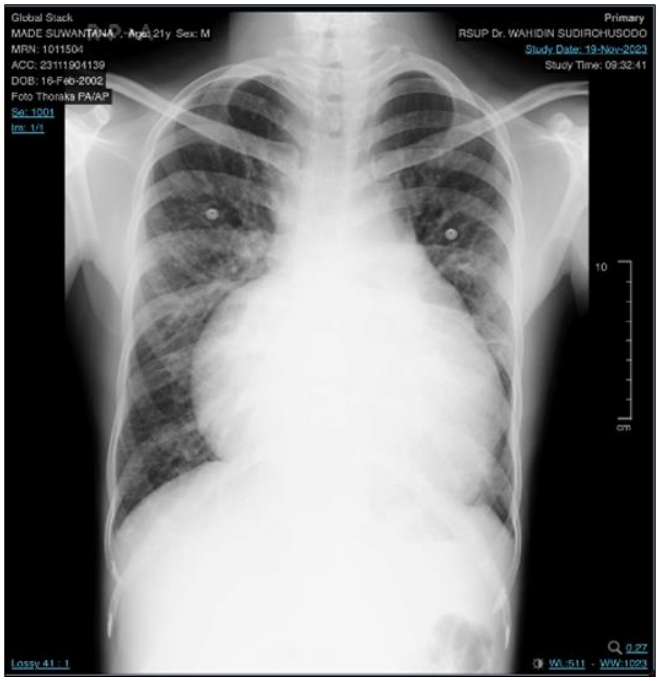
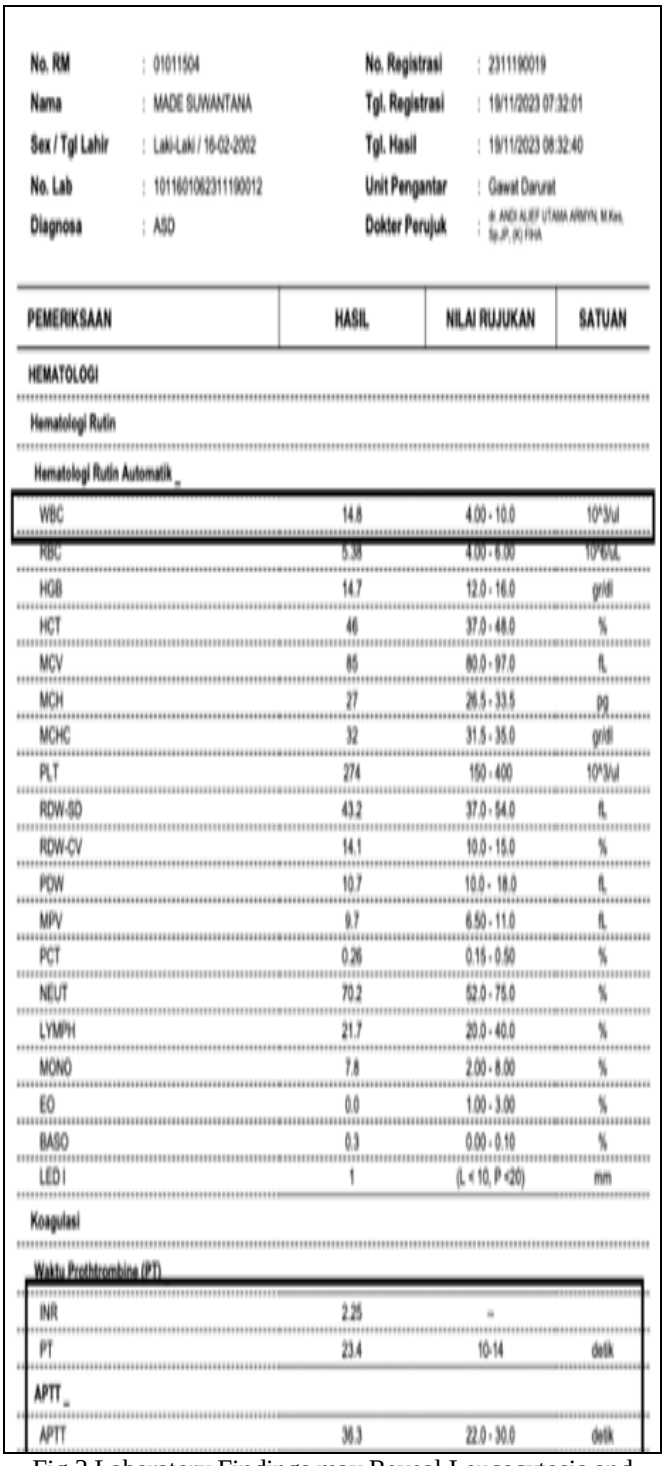



Fig 2 Chest Radiograph Showing Cardiomegaly (Cardiothoracic Ratio, 0.79) with a Left-to-Right Shunt Consistent with ASD, Pulmonary Oedema, and an ASD Occluder Device at CV T7-T9 Level

The chest X-ray reveals several findings suggestive of a heart condition: An enlarged heart. The ratio of the heart size to chest width (cardiothoracic ratio) is 0.79, which is higher than the normal value (typically below 0.5); This indicates abnormal blood flow from the left atrium (upper chamber) to the right atrium (upper right chamber) of the heart, possibly due to an atrial septal defect (ASD). An ASD is a hole in the wall separating these chambers; Fluid buildup in the lungs. This can be caused by the left-to-right shunt, where extra blood volume overwhelms the pumping capacity of the left ventricle, leading to backup and congestion in the lungs; The presence of a metallic device near the spine between the 7th and 9th thoracic vertebrae (T7-T9) suggests a previous procedure to close the ASD. This device plugs the hole in the atrial septum.

In summary, this chest X-ray indicates an enlarged heart, abnormal blood flow pattern likely due to a previously closed ASD, and fluid accumulation in the lungs, potentially caused by the earlier heart defect.



| | | | |
|---|---|---|---|
| No. RM | : 01011504 | No. Registrasi | : 2311190019 |
| Nama | : MADE SUWANTANA | Tgl. Registrasi | : 19/11/2023 07:32:01 |
| Sex / Tgl Lahir | : Laki-Laki / 16-02-2002 | Tgl. Hasil | : 19/11/2023 08:32:40 |
| No. Lab | : 10116010623111900l2 | Unit Pengantar | : Gawat Darurat |
| Diagnosa | : ASD | Dokter Perujuk | : dr. ANDI ALIEF UTAMA ARWYN, M.Kes, Sp.JP, (K) FIHA |

| PEMERIKSAAN | HASIL | NILAI RUJUKAN | SATUAN |
|---|---|---|---|
| HEMATOLOGI | | | |
| Hematologi Rutin | | | |
| Hematologi Rutin Automatik | | | |
| WBC | 14.8 | 4.00 - 10.0 | 10^3/ul |
| RBC | 5.38 | 4.00 - 6.00 | 10^6/uL |
| HGB | 14.7 | 12.0 - 16.0 | gr/dl |
| HCT | 46 | 37.0 - 48.0 | % |
| MCV | 86 | 80.0 - 97.0 | fL |
| MCH | 27 | 26.5 - 33.5 | pg |
| MCHC | 32 | 31.5 - 35.0 | gr/dl |
| PLT | 274 | 150 - 400 | 10^3/ul |
| RDW-SD | 43.2 | 37.0 - 54.0 | fL |
| RDW-CV | 14.1 | 10.0 - 15.0 | % |
| PDW | 10.7 | 10.0 - 18.0 | fL |
| MPV | 9.7 | 6.50 - 11.0 | fL |
| PCT | 0.26 | 0.15 - 0.50 | % |
| NEUT | 70.2 | 52.0 - 75.0 | % |
| LYMPH | 21.7 | 20.0 - 40.0 | % |
| MONO | 7.8 | 2.00 - 8.00 | % |
| EO | 0.0 | 1.00 - 3.00 | % |
| BASO | 0.3 | 0.00 - 0.10 | % |
| LED I | 1 | (L < 10, P <20) | mm |
| Koagulasi | | | |
| Waktu Prothrombine (PT) | | | |
| INR | 2.25 | - | |
| PT | 23.4 | 10-14 | detik |
| APTT | | | |
| APTT | 36.3 | 22.0 - 30.0 | detik |

Fig 3 Laboratory Findings may Reveal Leucocytosis and Prolonged Hemostatic Time

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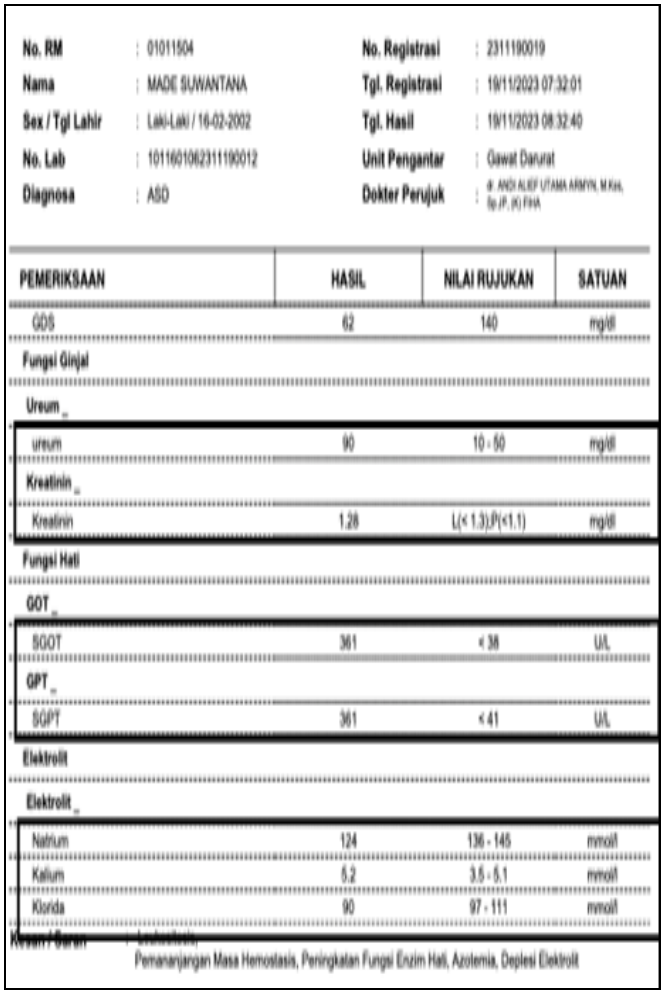

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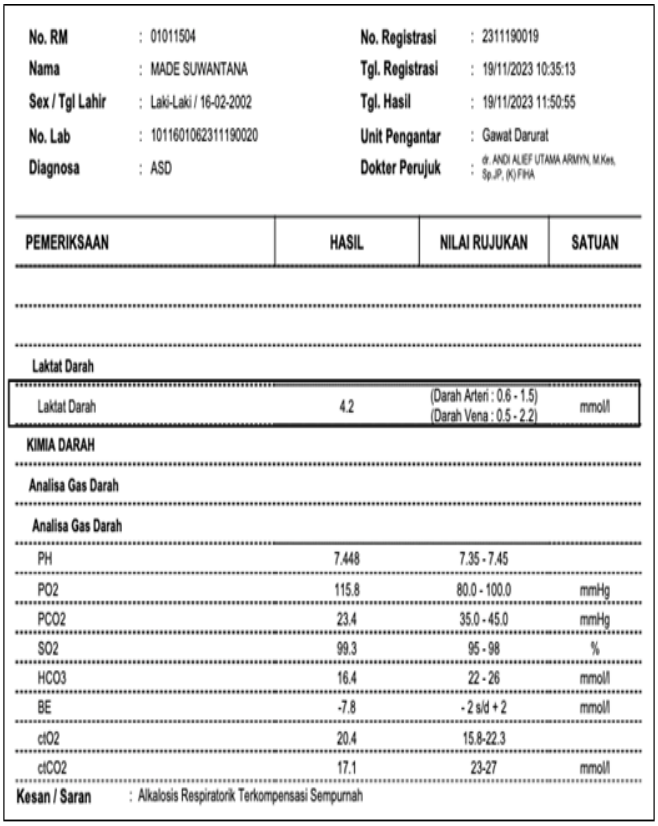
|---|---|---|---|
| No. RM | : 01011504 | No. Registrasi | : 2311190019 |
| Nama | : MADE SUWANTANA | Tgl. Registrasi | : 19/11/2023 07:32:01 |
| Sex / Tgl Lahir | : Laki-Laki / 16-02-2002 | Tgl. Hasil | : 19/11/2023 08:32:40 |
| No. Lab | : 10116010623111900l2 | Unit Pengantar | : Gawat Darurat |
| Diagnosa | : ASD | Dokter Perujuk | : dr. ANDI ALIEF UTAMA ARMYN, M.Kes, Sp.JP, (K) FIHA |

| PEMERIKSAAN | HASIL | NILAI RUJUKAN | SATUAN |
|---|---|---|---|
| GDS | 62 | 140 | mg/dl |
| **Fungsi Ginjal** | | | |
| **Ureum** | | | |
| ureum | 90 | 10 - 50 | mg/dl |
| **Kreatinin** | | | |
| Kreatinin | 1.28 | L(< 1.3);P(<1.1) | mg/dl |
| **Fungsi Hati** | | | |
| **GOT** | | | |
| SGOT | 361 | < 38 | U/L |
| **GPT** | | | |
| SGPT | 361 | < 41 | U/L |
| **Elektrolit** | | | |
| **Elektrolit** | | | |
| Natrium | 124 | 136 - 145 | mmol/l |
| Kalium | 5.2 | 3.5 - 5.1 | mmol/l |
| Klorida | 90 | 97 - 111 | mmol/l |
| Kesan / Saran | : Leukositosis, Pemanjangan Masa Hemostasis, Peningkatan Fungsi Enzim Hati, Azotemia, Deplesi Elektrolit | | |

Fig 4 Laboratory Findings may Reveal Azotemia, Elevated Liver Enzyme, and Electrolyte Depletion

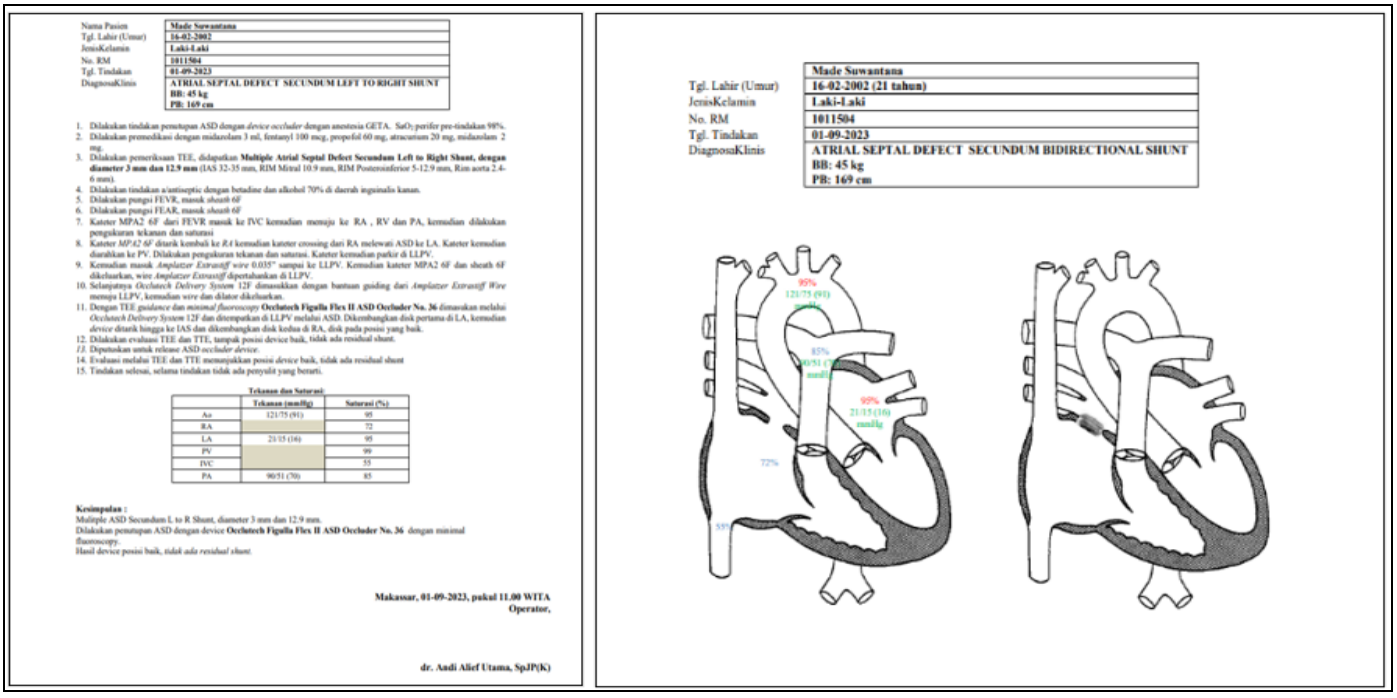


| | | | |
|---|---|---|---|
| No. RM | : 01011504 | No. Registrasi | : 2311190019 |
| Nama | : MADE SUWANTANA | Tgl. Registrasi | : 19/11/2023 10:35:13 |
| Sex / Tgl Lahir | : Laki-Laki / 16-02-2002 | Tgl. Hasil | : 19/11/2023 11:50:55 |
| No. Lab | : 10116010623111900l20 | Unit Pengantar | : Gawat Darurat |
| Diagnosa | : ASD | Dokter Perujuk | : dr. ANDI ALIEF UTAMA ARMYN, M.Kes, Sp.JP, (K) FIHA |

| PEMERIKSAAN | HASIL | NILAI RUJUKAN | SATUAN |
|---|---|---|---|
| **Laktat Darah** | | | |
| Laktat Darah | 4.2 | (Darah Arteri : 0.6 - 1.5) (Darah Vena : 0.5 - 2.2) | mmol/l |
| **KIMIA DARAH** | | | |
| **Analisa Gas Darah** | | | |
| **Analisa Gas Darah** | | | |
| PH | 7.448 | 7.35 - 7.45 | |
| PO2 | 115.8 | 80.0 - 100.0 | mmHg |
| PCO2 | 23.4 | 35.0 - 45.0 | mmHg |
| SO2 | 99.3 | 95 - 98 | % |
| HCO3 | 16.4 | 22 - 26 | mmol/l |
| BE | -7.8 | - 2 s/d + 2 | mmol/l |
| ctO2 | 20.4 | 15.8-22.3 | |
| ctCO2 | 17.1 | 23-27 | mmol/l |
| Kesan / Saran | : Alkalosis Respiratorik Terkompensasi Sempurnah | | |

Fig 5 Laboratory Findings may Reveal Completely Compensated Respiratory Acidosis

Previously, on 1 September 2023, the patient underwent ASD closure with an occluder device. Multiple ASD secundum L to R shunts were found, 3 mm and 12.9 mm in diameter, and ASD closure was performed with an Occlutech Figulla Flex II ASD Occluder No. 36 device under minimal fluoroscopy. The device was in good position with no residual shunt (Fig.6).



Nama Pasien : **Made Suwantana**
Tgl. Lahir (Umur) : **16-02-2002**
JenisKelamin : **Laki-Laki**
No. RM : **1011504**
Tgl. Tindakan : **01-09-2023**
DiagnosaKlinis : **ATRIAL SEPTAL DEFECT SECUNDUM LEFT TO RIGHT SHUNT**
**BB: 45 kg**
**PB: 169 cm**

1. Dilakukan tindakan penutupan ASD dengan *device occluder* dengan anestesia GETA. SaO2 perifer pre-tindakan 98%.
2. Dilakukan premedikasi dengan midazolam 3 ml, fentanyl 100 mcg, propofol 60 mg, atracurium 20 mg, midazolam 2 mg.
3. Dilakukan pemeriksaan TEE, didapatkan **Multiple Atrial Septal Defect Secundum Left to Right Shunt, dengan diameter *3* mm dan 12.9 mm** (IAS 32-35 mm, RIM Mitral 10.9 mm, RIM Posteroinferior 5-12.9 mm, Rim aorta 2.4-6 mm).
4. Dilakukan tindakan a/antiseptic dengan betadine dan alkohol 70% di daerah inguinalis kanan.
5. Dilakukan pungsi FEVR, masuk *sheath* 6F
6. Dilakukan pungsi FEAR, masuk *sheath* 6F
7. Kateter MPA2 6F dari FEVR masuk ke IVC kemudian menuju ke RA , RV dan PA, kemudian dilakukan pengukuran tekanan dan saturasi
8. Kateter *MP,62* 6F ditarik kembali ke *RA* kemudian kateter crossing dari RA melewati ASD ke LA. Kateter kemudian diarahkan ke PV. Dilakukan pengukuran tekanan dan saturasi. Kateter kemudian parkir di LLPV.
9. Kemudian masuk *Amplatzer Extrastiff wire* 0.035" sampai ke LLPV. Kemudian kateter MPA2 6F dan sheath 6F dikeluarkan, wire *Amplatzer Extrastiff* dipertahankan di LLPV.
10. Selanjutnya *Occlutech Delivery System* 12F dimasukkan dengan bantuan guiding dari *Amplatzer Extrastiff Wire* menuju LLPV, kemudian *wire* dan dilator dikeluarkan.
11. Dengan TEE *guidance* dan *minimal fluoroscopy* **Occlutech Figulla Flex II ASD Occluder No. 36** dimasukkan melalui *Occlutech Delivery System* 12F dan dimonpukan di LLPV melalui ASD. Dikembangkan disk pertama di LA, kemudian *device* ditarik hingga ke IAS dan dikembangkan disk kedua di RA, disk pada posisi yang baik.
12. Dilakukan evaluasi TEE dan TTE, tampak posisi device baik, tidak ada residual shunt.
13. Diputuskan untuk release ASD *occluder device*.
14. Evaluasi melalui TEE dan TTE memastikan posisi *device* baik, tidak ada residual shunt
15. Tindakan selesai, selama tindakan tidak ada penyulit yang berarti.

**Tekanan dan Saturasi:**

| | Tekanan (mmHg) | Saturasi (%) |
|---|---|---|
| Ao | 121/75 (91) | 95 |
| RA | | 72 |
| LA | 21/15 (16) | 95 |
| PV | | 99 |
| IVC | | 55 |
| PA | 90/51 (70) | 85 |

**Kesimpulan :**
Multiple ASD Secundum L to R Shunt, diameter 3 mm dan 12.9 mm.
Dilakukan penutupan ASD dengan device **Occlutech Figulla Flex II ASD Occluder No. 36** dengan minimal fluoroscopy.
Hasil device posisi baik, *tidak ada residual shunt.*

**Makassar, 01-09-2023, pukul 11.00 WITA**
**Operator,**

**dr. Andi Alief Utama, SpJP(K)**

Tgl. Lahir (Umur) : **16-02-2002 (21 tahun)**
JenisKelamin : **Laki-Laki**
No. RM : **1011504**
Tgl. Tindakan : **01-09-2023**
DiagnosaKlinis : **ATRIAL SEPTAL DEFECT SECUNDUM BIDIRECTIONAL SHUNT**
**BB: 45 kg**
**PB: 169 cm**

Fig 6 ASD Occluder Action Report Performed on 1 September 2023

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On echocardiography (Figure 7), the left atria (LA), right atria (RA) and right ventricle (RV) were dilated, and the left ventricle (LV) was D-shaped. The interatrial septal was intact, and there was a well-seated device with no residual shunt. The mitral valve had moderate regurgitation (VC 0.7 cm, mitral regurgitation ERO 0.2 cm2). The tricuspid valve had severe regurgitation (TR Vmax 436 cm/s, TR MaxPG 76 mmHg), the pulmonary valve had mild to moderate regurgitation (regurgitation jet width >1/3 right ventricular output tract, pulmonary regurgitation PHT 443 ms). The left ventricle had good systolic function with ejection fraction 66.8% (TEICH), grade III diastolic dysfunction (E/A >2; average E/e' 11.7), and there was a decline in systolic function in the right ventricle (TAPSE 1.12 cm). The pulmonary arteries were confluence and dilated (middle pulmonary artery 3.9cm, left pulmonary artery 1.6 cm, right pulmonary artery 1.3cm), with no patent ductus arteriosus. There was a mild pericardial effusion. Estimated right atrial pressure was 8 mmHg (1.8/1.2), and lung ultrasound may reveal multiple B-line <3 bilateral hemithorax, with no pleural effusion (-).

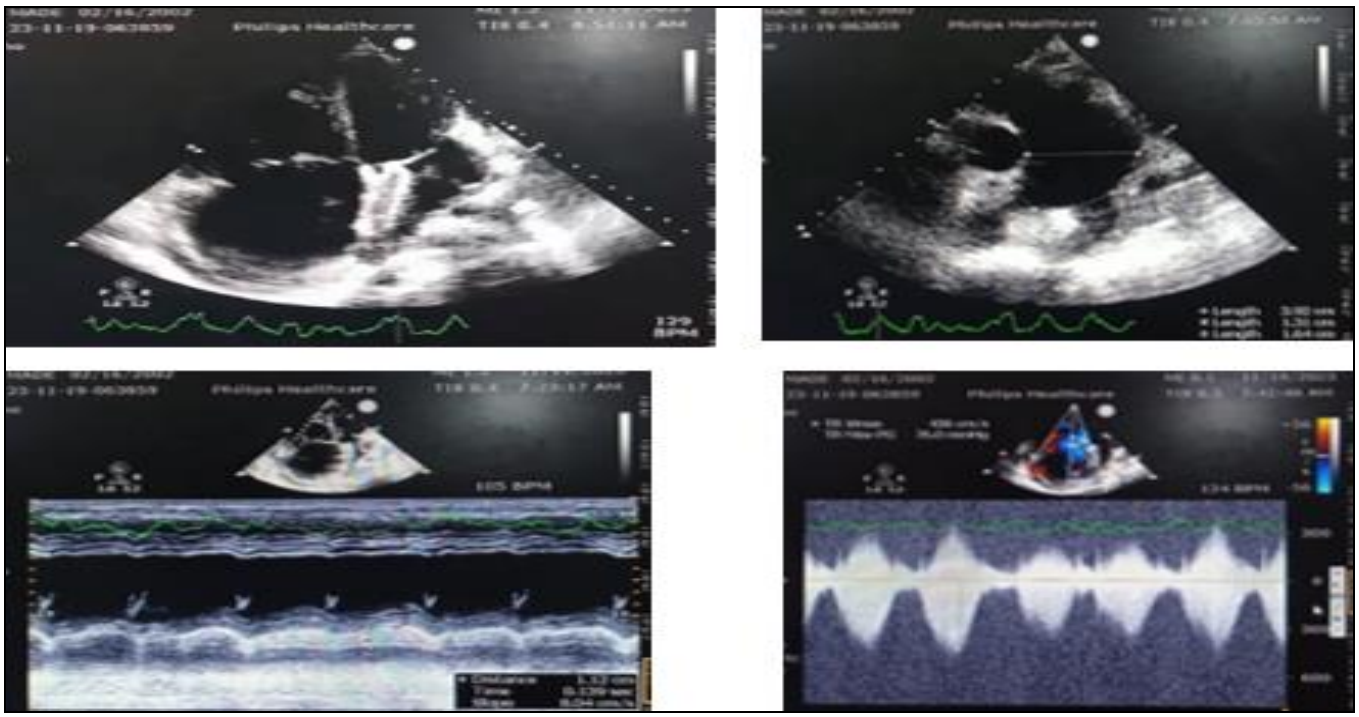

Fig 7 Echocardiography Findings

The patient was diagnosed with ASD Secundum Post Closure with Device (September 1th, 2023) and Pulmonary Hypertension Crisis. Patient was treated with digoxin, furosemide, milrinon, ceftriaxone, miniaspi, and plan for hemodynamic and vital sign monitoring, daily electrocardiography monitoring, urinalysis, and laboratory control for liver enzyme, ureum creatinine, and electrolyte. The patient was transferred to HCU. Four hours after receiving digoxin, the ECG revealed atrial flutter with fixed conduction, a regular heart rate of 115 bpm, right axis deviation (RAD) and right ventricle hypertrophy (RVH) (Figure 8).

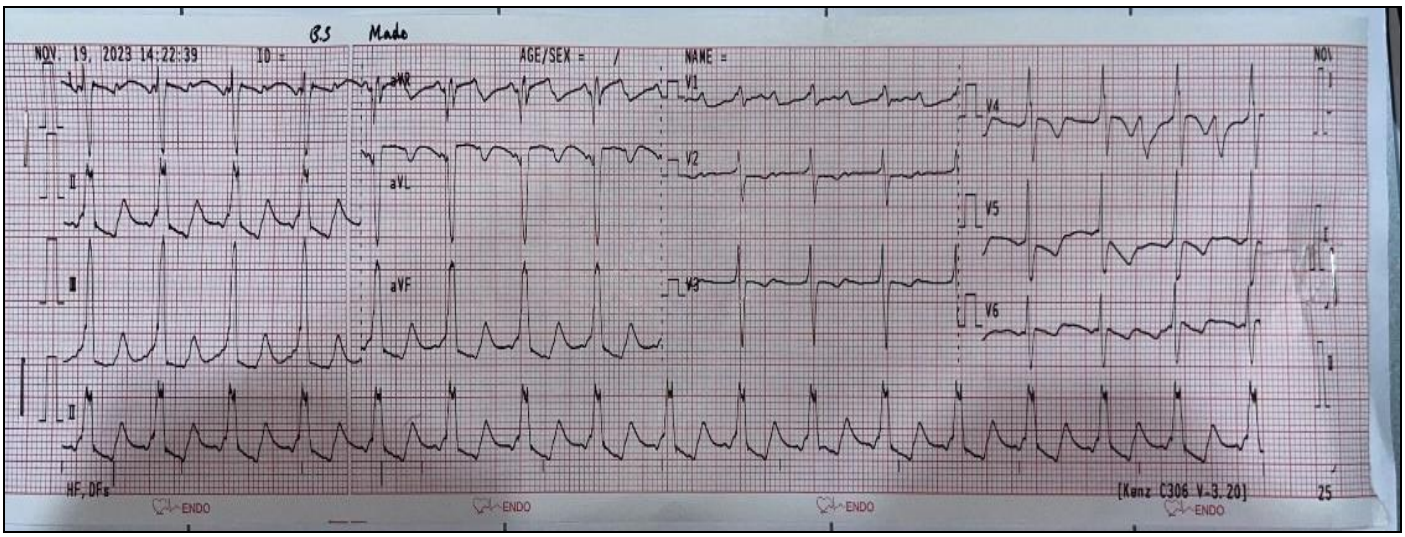


Fig 8 Electrocardiography 4 Hours after Digoxin. Atrial Flutter with Fixed Conduction, Heart Rate 115 Beats per Minute, Regular, Right Axis Deviation (RAD), and Right Ventricle Hypertrophy (RVH)

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The patient was then consulted in the electrophysiology subdivision on day 4 of treatment. By this time the dyspnoea had subsided and the patient could lie down at 30°. Pulse rate was 73 beats per minute, irregular, with different saturations in each limb (right arm 98%, left arm 97%, right leg 96% and left leg 96%). The patient's urine output was 1100cc/24h with a fluid balance of -390cc. A 12-lead electrocardiogram showed atrial flutter with variable conduction, average heartrate 70 beats per minute, irregular, right axis deviation (RAD), and right ventricle hypertrophy (RVH) (Figure 9).

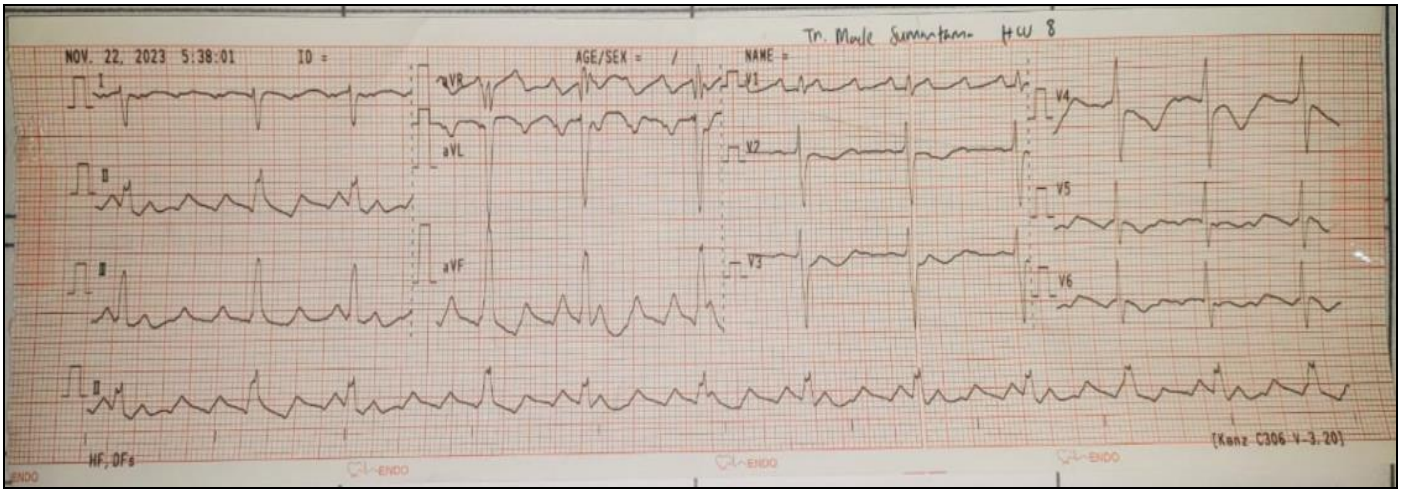
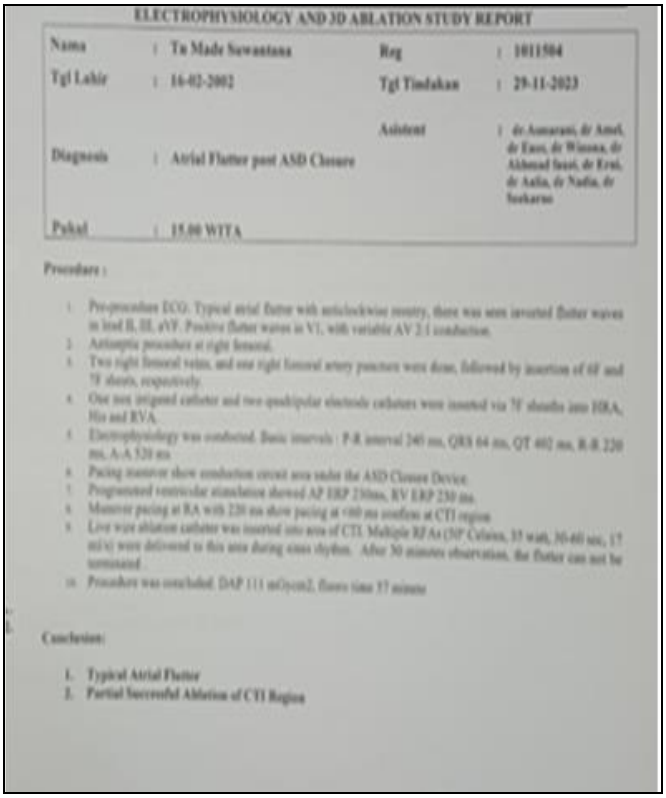



Fig 9 Atrial Flutter with Variable Conduction, Average Heartrate 70 Beats per Minute, Irregular, Right Axis Deviation (RAD), and Right Ventricle Hypertrophy (RVH).

The patient was scheduled for vital signs and haemodynamic monitoring, electrophysiology and 3D ablation. The electrophysiology and 3D ablation results concluded typical atrial flutter and partially successful ablation of CTI region. Figure 10 shows the electrophysiology and 3D ablation report.



ELECTROPHYSIOLOGY AND 3D ABLATION STUDY REPORT

| | | | |
|---|---|---|---|
| Nama | : Tn Made Suwantana | Reg | : 1011504 |
| Tgl Lahir | : 16-02-2002 | Tgl Tindakan | : 29-11-2023 |
| | | Asistent | : dr. Asmarani, dr. Astuti, dr. Enos, dr. Winona, dr. Akhmad Sanit, dr. Erni, dr. Aulia, dr. Nadia, dr. Sutkarno |
| Diagnosis | : Atrial Flutter post ASD Closure | | |
| Pukul | : 15.00 WITA | | |

Procedure :

1. Pre-procedure ECG: Typical atrial flutter with anticlockwise rotatory, there was seen inverted flutter waves in lead II, III, aVF. Positive flutter waves in V1, with variable AV 2:1 conduction.
2. Antiseptic procedure at right femoral.
3. Two right femoral veins, and one right femoral artery punctures were done, followed by insertion of 6F and 7F sheats, respectively.
4. One non irrigated catheter and two quadripolar electrode catheters were inserted via 7F sheaths into HRA, His and RVA.
5. Electrophysiology was conducted. Basic intervals : P-R interval 240 ms, QRS 64 ms, QT 402 ms, R-R 220 ms, A-A 520 ms
6. Pacing manover show conduction circuit area under the ASD Closure Device.
7. Programmed ventricular stimulation showed AP ERP 230ms, RV ERP 230 ms.
8. Manover pacing at RA with 220 ms show pacing at <60 ms confirm at CTI region.
9. Live wire ablation catheter was inserted into area of CTI. Multiple RFAs (30 Celsius, 35 watt, 30-60 sec, 17 ml/s) were delivered to this area during sinus rhythm. After 30 minutes observation, the flutter can not be terminated.
10. Procedure was concluded. DAP 111 mOsm/2, fluoro time 57 minute

Conclusion:

1. Typical Atrial Flutter
2. Partial Successful Ablation of CTI Region

Fig 10 An Electrophysiology and 3D Ablation Report Concluded that there was Typical Atrial Flutter and Partially Successful Ablation of CTI Region

## IV. DISCUSSION

The etiology of pulmonary hypertension is categorized into five distinct diagnostic groups: pulmonary arterial hypertension (PAH), pulmonary hypertension caused by left heart disease, pulmonary hypertension caused by lung disease or hypoxia, chronic thromboembolic pulmonary hypertension, and a multifactorial group (Authors/Task Force Members et al., 2009).

Pulmonary Arterial Hypertension (PAH) can be caused by various factors, but the key factor is that the rise in pressure in the lungs is caused by changes in the structure of the blood vessels in the pulmonary artery wall (Schermuly et al., 2011). In individuals diagnosed with Pulmonary Arterial Hypertension (PAH), the right ventricle is required to produce significantly elevated pressures compared to the average population. This is necessary to establish a pressure difference across the pulmonary circulation and provide sufficient blood flow. This results in the restructuring of the right ventricle while maintaining the function of the left ventricle. At first, there is an increase in the size of the right ventricle of the heart to compensate for a certain condition. Both the thickness of the right ventricular wall and the mass of the right ventricle increase dramatically. Individuals suffering with advanced pulmonary arterial hypertension (PAH) experience an enlargement in both the systolic and diastolic volumes of the right ventricle, along with a decrease in the right ventricular ejection fraction. Severe pulmonary arterial hypertension (PAH) results in a decrease in both systolic and diastolic function of the right ventricle, causing a large reduction in stroke volume. This ultimately leads to right heart failure (Temple, 2017).

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A sudden rise in pulmonary artery pressure (PAP) results in a temporary reduction in pulmonary blood flow (PBF) and blockage of the airways. This is partially caused by the constriction of tiny airways due to increased pressure in the pulmonary arterioles. Respiratory acidosis occurs as a consequence of elevated dead space ventilation and ventilation/perfusion (V/Q) mismatch. Simultaneously, there is an elevation in both right ventricular end-diastolic pressure (RVEDP) and volume (RVEDV). This hinders the supply of blood to the right coronary artery, which can result in a lack of oxygen to the heart muscle and the subsequent failure of the right ventricle. The interventricular septum is shifted towards the left, resulting in a decrease in the filling of the left ventricle (lower LV end-diastolic volume (LVEDV)) and cardiac output, which in turn leads to metabolic acidosis. This physiological process leads to circulatory collapse if not treated (Oishi and Fineman, 2016).

Pulmonary hypertension (PH) significantly raises the risk of illness and death in people with clinically significant atrial septal defect (ASD). Pulmonary hypertension (PH) is defined by the constriction and restructuring of the pulmonary arteries, resulting in the development of pulmonary vascular disease and subsequent right heart failure (HF). In cases where there is right-sided volume overload, it is advisable to intervene early and close the atrial septal defect (ASD). However, the impact of ASD closure on pulmonary hypertension (PH) is still a matter of debate. The prevalence of pulmonary hypertension (PH) in individuals with atrial septal defect (ASD) has been estimated to be between 10% and 20% according to studies by Akseer et al. (2022) and Stout et al. (2019).

A distinct subset of patients with severe pulmonary hypertension exhibit a concomitant secundum atrial septal defect (ASD), posing a challenge for the clinician in terms of treatment. The exact nature of the relationship between primary pulmonary hypertension and contemporaneous ASD in these individuals is uncertain. It is unclear whether the ASD has caused pulmonary hypertension gradually or if the patients already had primary pulmonary hypertension with an accompanying ASD. Pulmonary hypertension (PH) in individuals without surgical intervention for atrial septal defect (ASD) is linked to reduced functionality, atrial tachyarrhythmias, heart failure, and higher mortality rates. Despite the closure of the atrial septal defect (ASD), preoperative pulmonary hypertension (PH) continues to be a reliable indicator of mortality, heart failure, and arrhythmias. Pulmonary hypertension (PH) in the context of atrial septal defect (ASD) can result from many causes (Jain and Dalvi, 2018).

According to the general guidelines established by the American College of Cardiology and American Heart Association (Stout et al., 2019), it is not recommended and can be harmful to perform ASD closure in adults who have a net right to-left shunt at the ASD level, as well as systolic pulmonary artery to systolic systemic artery pressure and resistance ratios exceeding two-thirds. According to the guidelines, closure is typically recommended or considered appropriate when the ratio of pulmonary flow (Qp) to systemic flow (Qs) exceeds 1.5, with systolic pulmonary artery pressures being less than half of the systolic systemic pressures, and the ratio of pulmonary vascular resistance to systemic vascular resistance is less than one-third (Pan et al., 2020).

Pulmonary arterial hypertension (PAH) occurs in response to chronic volume overload of the pulmonary circulation caused by a left-to-right shunt that results in an alteration of the pulmonary vasculature. The haemodynamic definition of PH is a condition in which mPAP is > 20 mmHg at rest, and PH crisis is a clinical condition that occurs when mPAP exceeds MAP (Adriane et al., 2022; Nashat et al., 2018).

Acute PVR is a life-threatening event that can rapidly cause cardiorespiratory collapse. The initial assessment should address (Kaestner et al., 2016):

- Is the PH associated with systemic hypotension and/or hypoxaemia, low AVDO2, and thus most likely low cardiac output and tissue hypoxia?
- Are there any precipitating factors that might be responsible for elevated PVR and RV dysfunction (eg, infection, acidosis, arrhythmia, pericardial effusion)?
- Are there any other causes that could explain the symptoms of PHC and RV failure (eg, pneumothorax, pulmonary embolism)?

Following the initial clinical evaluation of vital signs, it is necessary to perform chest radiography and transthoracic echocardiography for proper therapy. Monitoring the hemodynamics and organ function, such as the brain, liver, kidney, and coagulation, is a crucial aspect of the routine and procedures in the Pediatric Intensive Care Unit (PICU) (Lammers et al., 2016).

Arterial and central venous lines should be set up for invasive monitoring in all patients with cardiopulmonary impairment who need vasopressor/inotropic medication. Invasive monitoring is recommended for patients who are at risk of developing low blood pressure due to targeted therapy for pulmonary hypertension, even if they are not receiving vasopressor or inotropic therapy (Lammers et al., 2016). Effective care of pulmonary hypertension (PH) following cardiac surgery involves two key components: (1) immediate intervention to address tissue hypoxia and severe acidosis, and (2) thorough assessment and treatment of right heart failure. Right ventricular failure is the primary cause of mortality in patients with pulmonary hypertension (PH). The function of the right ventricle (RV) is a significant factor that influences the occurrence of illness and death in this particular group of individuals. Right ventricular (RV) failure with low cardiac output and high RV filling pressures is common in PAH patients. PH crisis requires aggressive combination therapy and careful fluid management, inotropic and vasopressor control, maintenance of normal rhythm and pain control (Hoeper and Granton, 2011).

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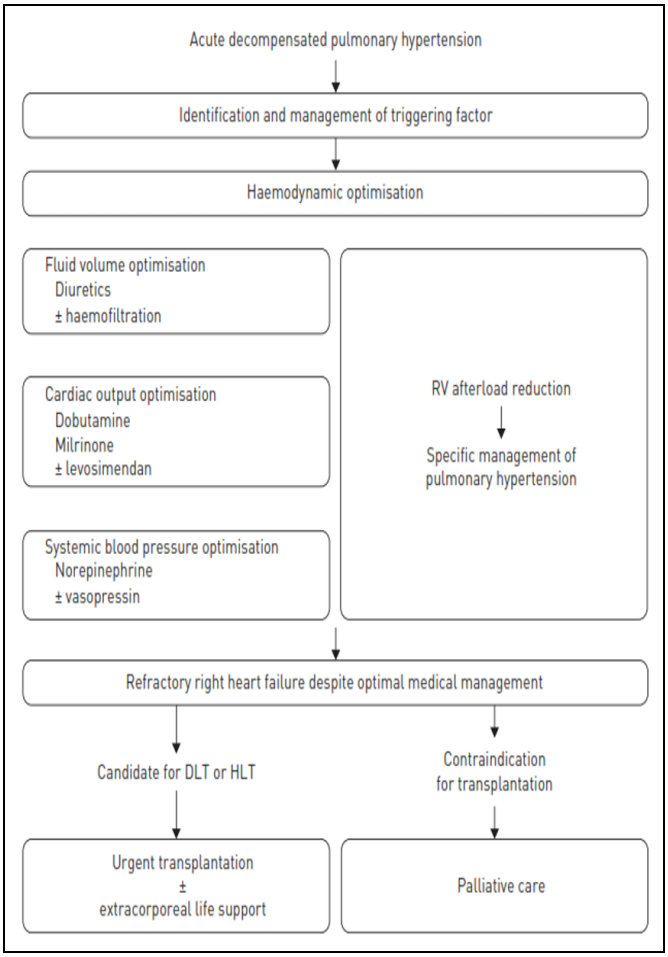


Fig 11 Management of Acute Decompensated Pulmonary Hypertension (Savale et al., 2017)

Supraventricular arrhythmias frequently occur in patients experiencing rapid decompensation. However, it can be challenging to ascertain if these arrhythmias are the cause or a consequence of right heart failure. Supraventricular arrhythmias mostly occur due to excessive blood volume and enlargement of the right ventricle, which is linked to a negative long-term outlook in cases of chronic pulmonary hypertension (Tongers et al., 2007). It decreases the amount of blood pumped with each heartbeat and might potentially lead to heart failure. Avoid the utilization of β-blockers and calcium channel inhibitors. These medicines adversely affect the contractility of the right ventricle. Medical cardioversion with amiodarone may be an option when the patient is receiving anticoagulation therapy. Radiofrequency ablation is a potential method to decrease the likelihood of atrial flutter recurring (Małaczyńska-Rajpold et al., 2016).

The level of blood filling in the right ventricle is strongly linked to the functioning of the heart in individuals with pulmonary hypertension (Sitbon et al., 2002). In cases of pulmonary hypertension, the impaired relaxation of the right ventricle leads to the accumulation of salt and water, which can become considerable during episodes of acute decompensated pulmonary hypertension. Intravenous diuretics are used to establish a state of negative fluid balance and optimize blood volume, resulting in a reduction in right ventricular preload. The objective of decreasing right ventricular preload is to diminish the connection between the right and left ventricles, thereby enhancing the function of the left ventricle during diastole. This approach also helps in minimizing tricuspid regurgitation and alleviating organ congestion, both of which are factors in the development of cardiorenal syndrome. One significant obstacle is the precise evaluation and tracking of right ventricular preload in these patients in order to ascertain the ideal amount of filling volume for each person. Both hypovolemia and hypervolemia have negative effects on heart function (Savale et al., 2017).

Diuretic therapy alone may not be enough to restore a condition of balance and recover from an episode of acute decompensation. In instances of profound right ventricular systolic failure, the utilization of inotropic drugs may be necessary to address a significantly compromised cardiac output. The primary objective of using inotropes is to enhance cardiac output and ventriculo-arterial coupling while avoiding an increase in right ventricular afterload (Kerbaul et al., 2004; Price et al., 2010). There are a limited number of clinical trials specifically assessing the effectiveness of inotropic drugs in the context of acute decompensated pulmonary hypertension. β1-adrenergic agonists continue to be the preferred inotropic drugs, despite preclinical studies showing reduced inotropic responses due to downregulation and desensitization of β1-receptors in right ventricular hypertrophy (Piao et al., 2012). Dobutamine is often started at a low dosage of 2.5 μg·kg−1·min−1 and gradually increased in patients who continue to show symptoms of poor cardiac output. Dobutamine demonstrated efficacy in enhancing ventriculo-arterial connection in an animal model of acute pulmonary artery blockage. Nevertheless, administering doses more than 10 μg·kg−1·min−1 can have harmful effects as it can cause a reduction in systemic arterial resistance without any improvement in cardiac output. Additional alternatives to dobutamine encompass more recent inotrope/vasodilators such milrinone or levosimendan. Milrinone is a medication that inhibits phosphodiesterase-3, leading to an increase in the force of contraction of the heart muscle and a decrease in the resistance faced by the left ventricle. Milrinone has demonstrated the ability to decrease pulmonary vascular resistance (PVR) in certain research investigations (Botha et al., 2009).

In patients with the most severe symptoms, the systemic vascular resistance is either low at admission or lowers following the use of diuretics, with or without inotropic drugs. This illness is commonly linked to the failure of many organs and the lack of blood supply to the right ventricle of the heart. In this scenario, it is imperative to utilize vasopressors like norepinephrine to reinstate a typical perfusion pressure. Norepinephrine has the potential to enhance both right ventricular function and the coupling between the right ventricle and the pulmonary artery, in addition to its impact on systemic blood pressure (Kerbaul et al., 2006). At lower doses, vasopressin can cause the blood vessels in the lungs to widen, which can help treat pulmonary hypertension. There have been documented occurrences of using vasopressin as a last resort treatment for this condition. When given in larger amounts, vasopressin can unexpectedly raise peripheral vascular resistance (PVR) and have negative effects on the

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heart (Price et al., 2010).Chronic right ventricular (RV) overload leads to the development of functional tricuspid regurgitation, which in turn causes an increase in right atrial (RA) pressure. Continued increase in right atrial pressure leads to gradual enlargement of the atrium and changes in its electrical properties. The remodeling of the right ventricle and right atrium, caused by prolonged pressure and volume overload, seems to create the arrhythmogenic substrate in individuals with pulmonary arterial hypertension (PAH). In addition to autonomic modulations, these patients may be more susceptible to developing atrial arrhythmias (AAs) (Medi et al., 2012; Rajdev et al., 2012).

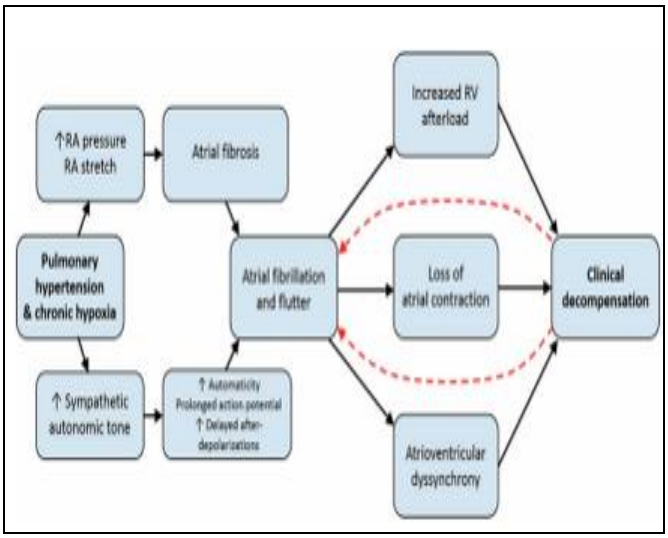


Fig 12 Mechanistic Pathways for the Pathogenesis of Atrial Arrhythmias in Pulmonary Hypertension and the Associated Pathophysiological Consequences (Wanamaker et al., 2018)

Patients with pulmonary arterial hypertension (PAH) frequently experience atrial tachyarrhythmias, including atrial flutter (AFL) and atrial fibrillation (AF). These conditions are commonly accompanied with a decline in heart function and clinical deterioration. Although patients with pulmonary arterial hypertension (PAH) face a higher risk of invasive procedures, right atrial catheter ablations are generally safe and highly effective for treating atrial arrhythmias. However, due to the severe enlargement/hypertrophy of the right atrium and the presence of multiple sites of origin/maintenance, these ablations may need to be extensive and repeated in PAH patients. Ventricular tachycardia is a rare condition, but relative bradycardia is a sign of something bad happening, commonly observed in cases of cardiopulmonary arrest. The reference is from Rajdev et al. (2012).

The European Society of Cardiology and European Respiratory Society guidelines for managing pulmonary hypertension propose rhythm control as the best technique for treating arrhythmias associated with this condition. Rhythm control options encompass pharmaceutical cardioversion using membrane antiarrhythmic medications, electrical cardioversion, and catheter ablation. Patients in this condition have a low tolerance for negative inotropic medications such as beta-blockers and calcium channel blockers. Due to their low tolerability and connection to negative results, the majority of observational studies on patients with PH and arrhythmias document an initial effort to control the rhythm (Galiè et al., 2016; Małaczyńska-Rajpold et al., 2016; Wanamaker et al., 2018).

Patients with pulmonary hypertension (PH) have been observed to benefit from the use of class III antiarrhythmic medications, such as amiodarone and sotalol (Wanamaker et al., 2018). Amiodarone may be advantageous in this context because it has a relatively neutral impact on cardiac contractility. Nevertheless, patients with pulmonary hypertension may not be able to utilize amiodarone for an extended period of time due to the presence of further lung disease and limited pulmonary function reserve. Amiodarone can elevate bosentan levels in patients receiving the endothelin receptor antagonist bosentan by inhibiting cytochrome P450 (CYP) 2C9. It is advisable to use caution when using Amiodarone (Dwyer and Kilpatrick, 2011).

Similar to patients without PH, catheter ablation is a first-line approach for the treatment of cavotricuspid isthmus (CTI)-dependent atrial flutter in patients with PH, despite the potential technical challenges of CTI ablation in PH patients with right-sided chamber dilatation. In PH patients undergoing catheter ablation for AF, mapping non-PV triggers, particularly in the RA, may improve outcomes when combined with traditional PVI-based approaches. Consider the risk of iatrogenic ASD with right-left shunt after transseptal access for left atrial ablation. This is particularly relevant in PH patients with baseline hypoxaemia and with the use of large bore transseptal sheaths (cryoballoons). Anaesthesia for catheter ablation in severe PH should be performed by providers with cardiac anaesthesia expertise (Wanamaker et al., 2018).

## V. CONCLUSION

Pulmonary hypertension can occur in cases of congenital heart defects, such as ASD. Chronic conditions can also trigger atrial arrhythmias which worsen the patient's condition. The operative management of ASD is closure of the ASD, but in some unique groups, this can lead to pulmonary hypertensive crisis after its closure. Therefore, ASD management needs to be done early to prevent other comorbidities that may accompany it.

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