Congenital Heart Disease

Tech Science Press

Doi:10.32604/chd.2025.064164

ARTICLE

Hemodynamic Profile Based on Right Heart Catheterization in Adult Acyanotic Congenital Heart Disease with Pulmonary Hypertension

Dina Anggraini[1], Kurnia Wahyudi[2], Melawati Hasan[3], Sri Endah Rahayuningsih[4,*] and Charlotte Johanna Cool[3]

[1]Faculty of Medicine, Padjadjaran University, Sumedang, 45363, Indonesia
[2]Department of Public Health, Faculty of Medicine, Padjadjaran University, Bandung, 40161, Indonesia
[3]Department of Cardiology, Dr.Hasan Sadikin General Hospital, Bandung, 40161, Indonesia
[4]Department of Pediatrics, Dr. Hasan Sadikin General Hospital, Bandung, 40161, Indonesia
*Corresponding Author: Sri Endah Rahayuningsih. Email: endah.perkani@gmail.com

Received: 07 February 2025; Accepted: 10 April 2025; Published: 30 April 2025

ABSTRACT: Background: Congenital heart disease (CHD) occurs in 9 out of 100 births and is the leading cause of birth defects, with acyanotic CHD being more common. The incidence of adult CHD is rising faster than pediatric CHD. Pulmonary hypertension is the most common complication in untreated CHD patients. Methods: This study is retrospective descriptive research based on medical record data and the results of right heart catheterization examinations in adult acyanotic CHD aged ≥18 years and free from other organ disorders. Results: A total of 103 patients met the inclusion criteria, the majority were young adults and predominantly female. The median height of 155.0 cm (IQR: 150.0–160.0), the mean weight was 49.6 kg ± 10.7 kg, and BSA of 1.4 m^2 ± 0.1 m^2. Most patients had a normal BMI status and were classified as WHO-FC II. Hemodynamic findings included a median PARI of 13.5 WU/m^2 (IQR: 4.5–29.6), RAP 7.0 mmHg (IQR: 5.0–11.0), mPAP 56.0 mmHg ± 23.5 mmHg, LVEDP 9.5 mmHg ± 3.9 mmHg, PVR 12.6 WU ± 11.0 WU, PVR/SVR 0.5 WU ± 0.4 WU, pulmonary artery saturation 74.0% ± 12.1%, aortic saturation 88.1% ± 7.8%, mixed venous saturation 59.0% ± 10.1%. Conclusions: This study provides comprehensive data on the demographic characteristics and hemodynamic parameters of patients with CHD and pulmonary hypertension. As a hospital-based registry, this study offers valuable insights into the clinical profile of adult CHD patients at Dr. Hasan Sadikin General Hospital, Bandung.

KEYWORDS: Adult congenital heart disease; acyanotic; pulmonary hypertension; hemodynamic

1 Introduction

Congenital Heart Disease (CHD) is the most common congenital abnormality affecting the heart, heart valves, and blood vessels [1]. It occurs in approximately 9 out of every 1.000 live births and remains the leading cause of all congenital defects [2]. Based on its impact on blood oxygen levels, CHD is classified into two major categories: acyanotic and cyanotic [3]. The severity of CHD varies, ranging from life-threatening abnormalities that hinder intrauterine survival to minor asymptomatic lesions that may result in delayed diagnosis or some cases remain undetected throughout life [4].

In 2019, an estimated 13.3 million people worldwide were living with CHD [1]. In 2017, global deaths attributed to CHD were estimated at 261.247 [5]. In Indonesia, the incidence of CHD is

approximately 43.200 cases per 4.8 million live births annually (9 per 1.000 live births) [6]. The global prevalence of adult CHD includes approximately 1.2 million cases in Europe and around 2 million cases in the United States [7]. Research indicates that acyanotic CHD is more common in adults than cyanotic CHD, with atrial septal defect (ASD) being the most prevalent at 60% followed by a ventricular septal defect (VSD) at 23% and patent ductus arteriosus (PDA) at 4%. Among cyanotic CHD cases in adults, tetralogy of Fallot (TOF) is the most prevalent, accounting for 8% [8].

Although CHD is present from birth, mild abnormalities may remain undetected for weeks, months, or even years and might only be diagnosed in adulthood [6]. The number of adult CHD cases is increasing at a faster rate compared to pediatric cases. This trend is largely because 90% of CHD conditions in children persist into adulthood, while the remaining 10% go undiagnosed during childhood [6]. If left untreated, CHD can lead to both cardiac and systemic complications [9].

One of the most severe complications of CHD is pulmonary hypertension, which occurs in approximately 10% of CHD patients [9]. Previous research has shown that among 1.012 patients, 411 (66.9%) adult CHD patients were confirmed to have pulmonary hypertension based on right heart catheterization. The diagnosis of CHD involves a combination of history taking, physical examination, chest X-ray evaluation, echocardiography, and right heart catheterization to assess hemodynamic status [9].

Given this background, research on the hemodynamic profile of adult patients with acyanotic CHD and pulmonary hypertension using right heart catheterization is crucial for evaluating the severity of hemodynamic disturbance. Therefore, this study aims to describe the hemodynamic profile of adult acyanotic CHD patients with pulmonary hypertension through right heart catheterization.

2 Methods

This study was a retrospective descriptive analysis based on medical records and right heart catheterization results of adult patients with acyanotic CHD and pulmonary hypertension at Dr. Hasan Sadikin General Hospital, Bandung, Indonesia from 2019 to 2023. The inclusion criteria comprised adult patients with acyanotic CHD who had undergone right heart catheterization and were diagnosed with pulmonary hypertension, defined as a mean pulmonary artery pressure (mPAP) >20 mmHg. Eligible participants were aged $\geq$18 years and had complete medical records. Using a total sampling method, 107 patients met the inclusion criteria.

The medical record data collected in this study included patient characteristics and hemodynamic parameters. Patient characteristics encompassed age, gender, height, weight, body surface area (BSA), and WHO-Functional Class (WHO-FC). Age was categorized according to Erikson's psychosocial development stage into young adulthood, middle-aged adulthood, and older adulthood. Gender was classified as male or female. Body mass index (BMI) was categorized based on the Asia Pacific classification: underweight (<18.5 kg/m^2), normal weight (18.5 kg/m^2–22.9 kg/m^2), overweight (23 kg/m^2–24.9 kg/m^2), and obesity ($\geq$25 kg/m^2). The WHO-FC was categorized into four classes: WHO-FC I, WHO-FC II, WHO-FC III, WHO-FC IV. Meanwhile, hemodynamic parameters obtained from right heart catheterization included the pulmonary artery resistance index (PARI), right atrial pressure (RAP), mPAP, left ventricular end-diastolic pressure (LVEDP), pulmonary vascular resistance (PVR), pulmonary vascular resistance/systemic vascular resistance ratio (PVR/SVR), pulmonary artery saturation, aortic saturation, and mixed venous saturation.

3 Results

According to the findings of this study, 103 adult patients with acyanotic CHD and pulmonary hypertension were identified at Dr. Hasan Sadikin General Hospital, Bandung, Indonesia. Between 2019 and 2023, with complete medical records and characteristics available for evaluation. Diagnosis of CHD was established using echocardiography, while pulmonary hypertension was confirmed through right heart catheterization [10,11]. In this assessment, a defining characteristic of CHD patients with pulmonary hypertension was an mPAP value exceeding 20 mmHg. Among the cases, ASD was the common condition, observed in 74 out of 103 patients (71.8%), followed by VSD in 17 patients (16.5%) and PDA in 12 patients (11.7%). Patient characteristics are presented in Table 1.

Table 1: Characteristics of Adult Acyanotic CHD with Pulmonary Hypertension.

Characteristics	Total (n = 103)
Height (cm) [median (IQR)][a]	155.0 (150.0–160.0)
Weight (kg) [mean ± SD][a]	49.6 ± 10.7
BSA (m^2) [mean ± SD][a]	1.4 ± 0.1
Age (years) [n(%)]	
Young Adult (18–40)	77 (74.8)
Middle-Age Adult (41–65)	25 (24.3)
Older (>65)	1 (1.0)
Gender [n(%)]	
Male	25 (24.3)
Female	78 (75.7)
BMI (kg/m^2) [n(%)][a]	
Underweight (<18.5)	37 (35.9)
Normal Weight (18.5–22.9)	44 (43.1)
Overweight (23–24.9)	12 (11.8)
Obesity (≥25)	9 (8.7)
WHO-FC [n(%)][b]	
I	1 (1.0)
II	85 (82.5)
III	7 (6.8)
IV	1 (1.0)

[a]: data of 102 patients; [b]: data of 94 patients; SD: standard deviation; BSA: body surface area; BMI: body mass index; WHO: World Health Organization.

The majority of adult patients with acyanotic CHD and pulmonary hypertension in this study had a median height of 155.0 cm, with varying ranges. The average BSA was 1.4 m^2, with a standard deviation of 0.1 m^2. The young adult age group had the highest prevalence among the categorized age groups. Additionally, female patients outnumbered male patients. Most patients had a normal BMI and were classified as WHO-FC II, indicating patients have mild symptoms with exertion, including dyspnea and fatigue, but no symptoms at rest.

The hemodynamic profile, assessed via right heart catheterization, yielded the following results (Table 2). Among 92 out of 103 patients, the median PARI value was 13.5 WU/m^2 (according to ESC, the normal PARI value is 3 WU/m^2–3.5 WU/m^2). Regarding RAP, 95 out of 103 patients had a median value above the normal range, at 7.00 mmHg (normal value according to ESC is 2 mmHg–6 mmHg). The mPAP value for all 103 patients was elevated, with an average of 56.0 mmHg ± 23.5 mmHg (normal mPAP value according to ESC is 8 mmHg–20 mmHg). Among the 55 out of 103 patients with available LVEDP values, the average

value was 9.5 mmHg ± 3.9 mmHg (normal mPAP value according to ESC is 5 mmHg–12 mmHg). The PVR was elevated in 91 out of 103 patients, with an average of 12.6 WU ± 11.0 WU (normal mPAP value according to ESC is 0.3 WU–2.0 WU). The PVR/SVR ratio was also increased in 74 out of 103 patients, averaging 0.5 WU ± 0.5 WU (normal value < 0.3 WU). Regarding oxygen saturation level, 88 out of 103 patients had an average pulmonary artery saturation of 74.0% ± 12.0% (normal pulmonary artery saturation value is 80%). Additionally, 85 out of 103 patients had an aortic saturation below normal with an average of 88.1% ± 7.8% (normal aortic saturation value is 96%–100%). The average mixed venous saturation value among 80 out of 103 patients was below the normal range at 59.3% ± 10.1% (normal mixed venous saturation value according to ESC is 65%–80%).

Table 2: Hemodynamic Profile Based on Right Heart Catheterization in Adult Acyanotic CHD with Pulmonary Hypertension.

Right Heart Catheterization Result	Total (n = 103)
PARI (WU/m^2) [median(IQR)][a]	13.5 (4.5–29.6)
RAP (mmHg) [median(IQR)][b]	7.0 (5.0–11.0)
mPAP (mmHg) [mean ± SD]	56.0 ± 23.5
LVEDP (mmHg) [mean ± SD][c]	9.5 ± 3.9
PVR (WU) [mean ± SD][d]	12.6 ± 11.0
PVR/SVR (WU) [mean ± SD][e]	0.5 ± 0.4
Pulmonary artery saturation (%) [mean ± SD][f]	74.0 ± 12.1
Aorta saturation (%) [mean ± SD][g]	88.1 ± 7.8
Mixed venous saturation (%) [mean ± SD][h]	59.0 ± 10.1

[a]: data of 92 patients; [b]: data of 95 patients; [c]: data of 55 patients; [d]: data of 91 patients; [e]: data of 74 patients; [f]: data of 88 patients; [g]: data of 85 patients; [h]: data of 80 patients; PARI: pulmonary artery resistance index; LVEDP: left ventricle end-diastolic pressure; RAP: right arterial pressure; SD: standard deviation; mPAP: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; SVR: systemic vascular resistance.

4 Discussion

According to data from the World Population Review, the average height of healthy individuals in Indonesia is 166 cm for males and 154 cm for females. However, in this study, height was not analyzed separately by sex, thus the relationship between body height on these conditions, has not been specifically analyzed [12]. A study by Daymont et al., found that children with CHD exhibited a significant deficit in weight, body length, and head circumference within a few weeks after birth compared to children without CHD [13]. This suggests that the presence of an unrepaired heart defect from birth may contribute to a height deficit in adult CHD patients compared to their healthy counterparts.

A study by Muljati et al., reported that the average body weight of healthy 18-year-old females is approximately 47.9 kg, while for males of the same age, it is around 54.2 kg [14]. However, in this study, height was not analyzed separately by sex, so the relationship between body weight on these conditions, has not been specifically analyzed. Despite these differences, a study by Brida et al., found a significant association between weight changes and prognosis in patients with complex CHD. Adult CHD patients who experienced weight loss had a significantly higher mortality rate compared to those without weight loss [15].

Based on the characteristics of body surface area (BSA) in adult patients with acyanotic CHD and pulmonary hypertension, the average BSA was 1.4 m^2 with a standard deviation of 0.1 m^2.

A study by Yang et al., reported similar findings, with a BSA of 1.49 ± 0.16 m^2 in adult CHD patients [16].

The majority of adult patients with acyanotic CHD and pulmonary hypertension are young adults (18–40 years). This is consistent with studies by Verheugt et al. and Rawat et al., which reported that most adult patients with acyanotic CHD fall within the age range of 20–30 years and 18 years, respectively [9,17]. In other words, these studies indicate that the young adult age group has a higher prevalence compared to other age categories. Most symptoms of CHD with pulmonary hypertension in adults are nonspecific. As a result, in older patients, initial evaluations are often focused on more common cardiovascular diseases, such as coronary artery disease. This may contribute to the high proportion of young adult CHD patients with pulmonary hypertension referred to our hospital, as pulmonary hypertension may have gone undetected or overlooked during prior evaluations at other healthcare facilities.

Based on gender characteristics, adult patients with acyanotic CHD are predominantly female. This finding aligns with a cohort study conducted in Taiwan by Chiu et al. which reported that CHD with pulmonary hypertension was more commonly observed in females than in males [18]. Similarly, a study in Japan by Yao et al., found that female patients were more prevalent in certain types of CHD, including ASD, VSD, and PDA [19]. Sex hormones and developmental factors have been hypothesized as potential contributors to these gender differences in congenital anomalies [20].

In this study, most patients had a normal BMI, consistent with findings by Zaqout et al., which indicated that normal or underweight BMI is more commonly observed in adult CHD patients. Additionally, BMI status in adult CHD patients has been linked to the severity of the cardiac defect [21]. However, since this study did not assess defect severity, the relationship between BMI and defect severity could not be directly evaluated. Furthermore, a study by Brida et al., found that a higher BMI is associated with a lower mortality rate in adult CHD patients [15].

In terms of WHO-FC characteristics, adult patients with acyanotic CHD are predominantly classified as WHO-FC II. This finding is consistent with a study by Kücükoglu et al., which reported that WHO-FC II status was the most common classification among CHD patients with pulmonary hypertension [22]. Basically, WHO-FC status is recognized as one of the strongest predictors of survival, both at diagnosis and during follow-up. A worsening WHO-FC classification is a key indicator of disease progression in pulmonary hypertension [10]. As widely known, higher WHO-FC classifications are associated with an increased one-year mortality risk in CHD patients with pulmonary hypertension. Therefore, the majority of patients at our hospital classified as WHO-FC II are considered to have a lower estimated one-year mortality risk.

This study, along with the study conducted by Dinarti et al., both reported abnormal PARI values, with a median value of 33 WU/m^2 and a minimum-maximum value of 1.6 WU/m^2–11.6 WU/m^2. PARI is essentially the PVR value adjusted for a person's BSA, providing a more standardized assessment of PVR. The PVR value itself is influenced by several factors, including the size of intracardiac defects. Consequently, an individual's PARI value is also indirectly affected by the size of the heart defect [23]. The variation in PARI values between this study and the previous study by Dinarti et al., may be attributed to differences in defect size among adult CHD patients [9]. However, since defect size was not analyzed in this study, its direct relationship with PARI values could not be evaluated.

The RAP values in this study were above normal. Similarly, a study by Dinarti et al., also reported abnormal RAP values in adult CHD patients with pulmonary hypertension, with a median value of 9.0 mmHg and a range of 6.0 mmHg~13.0 mmHg [9]. The RAP is one of the hemodynamic parameters for assessing prognosis, as values below 8 mmHg are associated with a lower risk of mortality within one year. A recent study from France identified WHO-FC, six-minute walk distance (6 MWD), RAP, and stroke volume index (SVI) as independent predictors of prognosis in pulmonary hypertension associated with CHD [10]. Based on these findings, adult acyanotic CHD patients with pulmonary hypertension in this study are estimated to have a low 1-year mortality risk.

According to the latest clinical classification from the 6th World Symposium of Pulmonary Hypertension (WSPH), an individual is diagnosed with pulmonary hypertension when the mPAP value exceeds 20 mmHg [24]. The findings of this study, along with previous studies by Chiu et al., and Idress et al., confirm mPAP values consistent with the definition of pulmonary hypertension [18,25]. An increase in mPAP significantly raises the risk of mortality, particularly when the value exceeds 25 mmHg [23]. Although the average mPAP value in this study's patients was higher, the estimated 1-year mortality risk remains low based on WHO-FC. This is because WHO-FC remains the strongest predictor of disease severity and clinical outcomes, surpassing mPAP in prognostic significance.

The LVEDP values are used to determine the type of pulmonary hypertension. According to ESC guidelines, a PAWP value of $\leq$15 mmHg indicates pre-capillary pulmonary hypertension, which includes pulmonary hypertension caused by CHD (LVEDP values are often considered equivalent to PAWP) [10,23]. The findings of this study support this classification, as the average LVEDP value of $\leq$15 mmHg in study participants indicates they fall into the pre-capillary pulmonary hypertension category. Furthermore, the value obtained in this study serves as an indicator that pulmonary hypertension in these patients is entirely attributed to CHD, with no indication of left heart disease involvement or a mixed etiology as contributing factors.

The findings of this study are consistent with research conducted by Idress et al., which also reported abnormally high PVR values [25]. This aligns with the natural progression of pulmonary hypertension, where PVR increases due to remodeling of the pulmonary blood vessels [26]. PVR also serves as a crucial indicator for assessing the safety of defect closure in CHD patients, with a PVR <5 WU generally considered a safer threshold. In this study, with an average PVR $\geq$ 5 WU, defect closure in these patients is associated with a higher risk of poor outcomes. Therefore, it is recommended that patients first undergo treatment for pulmonary hypertension, followed by a reevaluation of hemodynamics, and consider defect closure only if the PVR value falls below 5 WU [26].

Studies by Kücükoglu et al. and Amsallem et al., reported similar findings regarding the PVR/SVR ratio, with the values exceeding normal levels [22,27]. An elevated PVR/SVR ratio indicates increased resistance in the pulmonary vasculature relative to systemic circulation. Under normal conditions, SVR is higher than PVR. However, when PVR surpasses SVR, the direction of shunting from the left heart to the right heart can reserve, leading to cyanosis a condition characterized by bluish discoloration of the skin and mucous membrane due to low oxygen levels in the blood [23]. Based on the PVR/SVR ratio values observed at our hospital, the majority of patients exhibited elevated ratios, suggesting a higher risk of progression toward more severe forms of cyanotic CHD.

The pulmonary artery saturation values in this study were below normal. Similarly, a study by Thomas et al., reported an average pulmonary artery saturation of 68% ± 7%, which was also lower than normal [28]. In CHD, pulmonary artery saturation is commonly used to detect the presence of a left-to-right shunt. However, in this study, patients had already developed pulmonary hypertension, with pulmonary artery pressure exceeding normal levels. This likely led to a reduction in the left-to-right shunt, bringing pulmonary artery saturation values closer to normal.

The aortic saturation characteristics in this study showed an average value of 88.1% ± 7.8% whereas the normal range for aortic saturation is 96%–100%. This finding aligns with a study by Thomas et al., which reported an average aortic saturation value of 88% ± 5% [28]. In our study, aortic saturation values below the normal range indicate the mixing of deoxygenated blood from the right side to the left side of the heart. This finding is also consistent with the previously discussed increase in the PVR/SVR ratio. However, further evaluation using more specific diagnostic tests is needed to confirm the direction of the shunt in the heart defect.

Mixed venous saturation values serve as an indicator of the presence of an intracardiac shunt. A mixed venous oxygen saturation level exceeding 75% suggests abnormal intracardiac blood flow, potentially indicating a left-to-right shunt [10]. However, in this study, the mixed venous oxygen saturation values were found to be <75%. This finding is consistent with the principle of pulmonary artery saturation, where CHD patients in this study had progressed to pulmonary hypertension. The elevated pulmonary pressure likely reduced left-to-right shunting. This perspective is supported by a study conducted by Mueller-Mottet et al., which found that CHD patients with pulmonary hypertension had mixed venous saturation values below 75% [29].

This study is a single-center, hospital based, and therefore, its findings cannot yet be generalized as the hemodynamic profile of adult acyanotic CHD patients with pulmonary hypertension across West Java or larger regions. Dr. Hasan Sadikin General Hospital is not the sole referral center for adult CHD patients in West Java, as some patients also seek treatment at private hospitals. Consequently, the study sample may not fully represent the boarder population of adult CHD patients in the region. Thus, future studies with multiple cohorts or institutions are warranted to validate our findings.

5 Conclusions

This study documents data on patients with CHD and pulmonary hypertension, presenting both demographic characteristics and hemodynamic parameters. As a hospital-based registry, it provides a comprehensive overview of the clinical profile of adult CHD patients with pulmonary hypertension at Dr. Hasan Sadikin General Hospital, Bandung. Further cohort studies on routine right heart catheterization in pediatric CHD are needed to identify the risk of early pulmonary hypertension development, allowing for earlier clinical interventions to reduce mortality rates and improve the quality of life of patients.

Acknowledgement: We would like to express our gratitude to Dr. Achmad Fitrah Khalid, Sp.JP., Dr. Fatihatul Firdaus, and Dr. Norman Sukmadi from the Department of Cardiology, Dr. Hasan Sadikin General Hospital, Bandung, 40161, Indonesia for their guidance and support in data collection for this research.

Funding Statement: The authors received no specific funding for this study.

Author Contributions: Dina Anggraini: Investigation, Writing of the main manuscript, Statistical analysis. Charlotte Johanna Cool: Conceptualization, Review of the main manuscript. Sri Endah Rahayuningsih: Conceptualization, Review of the main manuscript. Melawati Hasan: Review of the study concept. Kurnia Wahyudi: Review of the methodology. All authors reviewed the results and approved the final version of the manuscript.

Availability of Data and Materials: The article contains all of the tables. Please be aware, though that institutional safety protocols and policies prevent direct access to raw databases and further information. Researchers can send a formal request to our university by letter if they are interested in accessing the database.

Ethics Approval: This study has been approved by the Ethics Committee of Padjadjaran University (approval number 181/UN6.KEP/EC/2024 of February 23, 2024) and has been approved by the ethics committee of Dr. Hasan Sadikin General Hospital Bandung (approval number DP.04.03/D.XIV.4.4/450/2024 of March 26, 2024).

Conflicts of Interest: The authors declare no conflicts of interest to report regarding the present study.

References

1. Tsao CW, Aday AW, Almarzooq ZI, Alonso A, Beaton AZ, Bittencourt MS, et al. Heart disease and stroke statistics—2022 update: a report from the American heart association. Circulation. 2022;145(8):e153–639. [CrossRef].
2. Wing EJ, Schiffman FJ. Cecil Essentials of Medicine. 10th ed. Amsterdam, The Netherlands: Elsevier; 2022.
3. Kumala K, Yantie NPVK, Hartawan NB. Karakteristik Penyakit Jantung Bawaan Asianotik Tipe Isolateed dan Manifestasi Klinis Dini Pada Pasien Anak di Rumah Sakit Umum Pusat Sanglah. E-J Med Udayana. 2018;7(10). (In Indonesia).
4. Kumar V, Abbas AK, Aster JC. Robbins Basic Pathology. 10th ed. New York, NY, USA: Elsevier; 2018.
5. Zimmerman MS, Smith AGC, Sable CA, Echko MM, Wilner LB, Olsen HE, et al. Global, regional, and national burden of congenital heart disease, 1990–2017: a systematic analysis for the global burden of disease study 2017. Lancet Child Adolesc Health. 2020;4(3):185–200. [CrossRef].
6. Marwali EM, Purnama Y, Roebiono PS. Modalitas deteksi dini penyakit jantung bawaan di pelayanan kesehatan primer. J Indones Med Assoc. 2021;71(2):100–9. (In Indonesian). [CrossRef].
7. Weale J, Kelleher AA. Adult congenital heart disease. Anaesth Intensive Care Med. 2024;25(4):249–55. [CrossRef].
8. Ismail MT, Hidayati F, Krisdinarti L, Noormanto , Nugroho S, Wahab AS. Epidemiological profile of congenital heart disease in a national referral hospital. J Acta Cardiol Indones. 2015;1(2):66–71.
9. Dinarti LK, Hartopo AB, Kusuma AD, Satwiko MG, Hadwiono MR, Pradana AD, et al. The congenital heart disease in adult and pulmonary hypertension (COHARD-PH) registry: a descriptive study from single-center hospital registry of adult congenital heart disease and pulmonary hypertension in Indonesia. BMC Cardiovasc Disord. 2020;20:1–11. [CrossRef].
10. Humbert M, Kovacs G, Hoeper MM, Badagliacca R, Berger RMF, Brida M, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension: developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European society of cardiology (ESC) and the European Respiratory society (ERS). endorsed by the international society for heart and lung transplantation (ISHLT) and the European reference network on rare respiratory diseases (ERN-LUNG). Eur Heart J. 2022;43(38):3618–731. [CrossRef].
11. Myers PO, Tissot C, Beghetti M. Assessment of operability of patients with pulmonary arterial hypertension associated with congenital heart disease—do we have the good tools to predict success? Circ J. 2014;78(1):4–11. [CrossRef].

12. World Population Review. Average Height by Country 2025 [Internet]. [cited 2025 Feb 6]. Available from: https://worldpopulationreview.com/country-rankings/average-height-by-country.
13. Daymont C, Neal A, Prosnitz A, Cohen MS. Growth in children with congenital heart disease. Pediatrics. 2013;131(1):e236–42. [CrossRef].
14. Muljati S, Triwinarto A, Utami N, Hermina H. Gambaran median tinggi badan dan berat badan menurut kelompok umur pada penduduk indonesia yang sehat berdasarkan hasil riskesdas 2013. Penelit Gizi dan Makanan. J Nutr Food Res. 2017;39(2):137–44. (In Indonesian). [CrossRef].
15. Brida M, Dimopoulos K, Kempny A, Liodakis E, Alonso-Gonzalez R, Swan L, et al. Body mass index in adult congenital heart disease. Heart. 2017;103(16):1250–7. [CrossRef].
16. Yang ZY, Li HZ, Xie NS, Zhou Y, Luo DL, Fei HW, et al. Model for screening adult congenital heart disease surgery eligibility with echocardiography parameters. J Heart Lung Transplant. 2022;41(12):1831–8.
17. Rawat A, Belwal CM, Gururani K. Prevalence of adult congenital cardiac disease: report from a tertiary care center. Pak Heart J. 2023;56(3):528–36.
18. Chiu SN, Lu CW, Lin MT, Chen CA, Wu MH, Wang JK. Pulmonary hypertension in adult congenital heart disease in Asia: a distinctive feature of complex congenital heart disease. J Am Heart Assoc. 2022;11(7):e022596. [CrossRef].
19. Yao A, Inuzuka R, Mizuno A, Iwano H, Tatebe S, Tsukamoto Y, et al. Status of adult outpatients with congenital heart disease in Japan: The Japanese network of cardiovascular departments for adult congenital heart disease registry. J Cardiol. 2022;80(6):525–31. [CrossRef].
20. Pugnaloni F, Felici A, Corno AF, Marino B, Versacci P, Putotto C. Gender differences in congenital heart defects: a narrative review. Transl Pediatr. 2023;12(9):1753–64. [CrossRef].
21. Zaqout M, Vandekerckhove K, Michels N, Demulier L, Bove T, François K, et al. Body mass index in adults with congenital heart disease. Congenit Heart Dis. 2019;14(3):479–86. [CrossRef].
22. Kücükoglu SM, Kaymaz C, Alehan D, Kula S, Akcevin A, Celiker A, et al. Pulmonary arterial hypertension associated with congenital heart disease: lessons learnt from the large Turkish nationwide registry (THALES). Pulm Circ. 2021;11(3): 20458940211024206. [CrossRef].
23. Libby P, Bonow RO, Mann DL, Tomaselli GF, Bhatt DL, Solomon SD. Braunwald's Heart Disease a Textbook of Cardiovascular Medicine. 12th ed. Braunwald E, editor. Philadelphia, PA, USA: Elsevier; 2022.
24. Jone PN, Ivy DD, Hauck A, Karamlou T, Truong U, Coleman RD, et al. Pulmonary hypertension in congenital heart disease: a scientific statement from the American heart association. Circ Hear Fail. 2023;16(7):e00080. [CrossRef].
25. Idrees MM, Al-Najashi K, Khan A, Al-Dammas S, Al-Awwad H, Batubara E, et al. Pulmonary arterial hypertension in Saudi Arabia: patients' clinical and physiological characteristics and hemodynamic parameters. a single center experience. Ann Thorac Med. 2014;9(4):209–15. [CrossRef].
26. Baumgartner H, de Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al. 2020 ESC guidelines for the management of adult congenital heart disease: the task force for the management of adult congenital heart disease of the European society of cardiology (ESC). endorsed by: association for European paediatric and congenital cardiology (AEPC), international society for adult congenital heart disease (ISACHD). Eur Heart J. 2021;42(6):563–645. [CrossRef].
27. Amsallem M, Bagherzadeh SP, Boulate D, Sweatt AJ, Kudelko KT, Sung YK, et al. Hemodynamic trajectories and outcomes in patients with pulmonary arterial hypertension. Pulm Circ. 2020;10(4):2045894020941343. [CrossRef].
28. Thomas IC, Glassner-Kolmin C, Gomberg-Maitland M. Long-term effects of continuous prostacyclin therapy in adults with pulmonary hypertension associated with congenital heart disease. Int J Cardiol. 2013;168(4):4117–21. [CrossRef].
29. Mueller-Mottet S, Stricker H, Domeninghetti G, Azzola A, Geiser T, Schwerzmann M, et al. Long-term data from the swiss pulmonary hypertension registry. Respiration. 2015;89(2):127–40. [CrossRef].