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Original Research Article

ANALYSIS OF INCIDENCE OF IDIOPATHIC PULMONARY HYPERTENSION IN CHILDREN: AN INSTITUTIONAL BASED STUDY

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ABSTRACT

Background: Paediatric pulmonary hypertension (PH) is a heterogeneous disorder associated with significant morbidity and mortality if untreated. Idiopathic pulmonary arterial hypertension (IPAH), a rare subgroup of PH, is characterized by elevated mean pulmonary artery pressure (mPAP >20 mmHg), pulmonary arterial wedge pressure (PAWP ≤15 mmHg), and pulmonary vascular resistance index (PVRi >3 WU·m²). In children, IPAH often presents early and carries a poor prognosis if not detected promptly. This study aimed to determine the incidence, demographic distribution, and clinical presentation of IPAH in children.

Materials and Methods: A prospective, observational, institution-based study was conducted in the Department of Pediatrics, Parul Institute of Medical Sciences and Research, Vadodara, India, over six months (October 2024–March 2025). A total of 1,000 children aged ≤10 years were screened. Diagnosis of IPAH was based on echocardiographic evaluation and, where indicated, right heart catheterization. Children with PH secondary to congenital heart disease, lung disease, left heart disease, or systemic disorders were excluded. Demographic details, clinical features, and echocardiographic findings were analyzed using descriptive statistics.

Results: Of the 1,000 children evaluated, 50 (5%) were diagnosed with IPAH. Males constituted 60% (n=30) and females 40% (n=20), with a male-to-female ratio of 1.5:1. The majority of cases occurred in the 1–5 year age group (80%). Common clinical presentations included shortness of breath (80%), chest pain (80%), functional dyspnea (60%), and peripheral edema (60%). Syncope, hepatomegaly, and ascites were noted in 40% of cases, while cyanosis and heart murmurs were present in 20%. Palpitations were absent in all cases.

Conclusion: The study revealed a 5% incidence of IPAH among children, with early childhood predominance and a slight male bias. Dyspnea and chest pain were the leading clinical features. Early echocardiographic evaluation in children with unexplained dyspnea or right heart strain is essential for timely diagnosis, enabling prompt intervention and improved outcomes.

Keywords: Pediatric pulmonary hypertension, Idiopathic pulmonary arterial hypertension, Echocardiography, Dyspnea, Children.

INTRODUCTION

Paediatric pulmonary hypertension (PH) represents a heterogeneous illness that is responsible for high morbidity and mortality if left without treatment.[1] In fact it is represented by the harmful combination of increased mean pulmonary artery pressure (mPAP) at rest >20 mmHg, pulmonary arterial wedge pressure (PAWP) ≤15 mmHg, and pulmonary vascular resistance index (PVRi) >3 WU·m^2 in patients with biventricular physiology being subjected to right heart catheterisation (RHC).[2] Pediatric pulmonary hypertension is intrinsically linked to lung growth and development

in the younger child.[3] The onset of pulmonary vascular injury in the younger child may allow the possibility of greater reversal of pulmonary vascular disease, particularly in bronchopulmonary dysplasia (BPD) and other lung diseases of childhood.[4] Idiopathic pulmonary arterial hypertension (IPAH) is a subtype of PAH rarely seen in paediatrics. An exact underlying risk factor is unknown. It is classified in group I PH, together with PH triggered by congenital heart disease.[5] IPAH can manifest in newborns or infants in the form of persistent pulmonary hypertension of the newborn (PPHN).[6] The present study was conducted to analyze incidence of idiopathic pulmonary hypertension in children.

MATERIALS AND METHODS

Study Design and Setting

This was a prospective, observational, institution-based study conducted in the Department of Pediatrics, Parul Institute of Medical Sciences and Research, Vadodara, Gujarat, India. The study was carried out over a period of six months (October 2024 to March 2025) after obtaining prior approval from the Institutional Ethics Committee.

Study Population

Children attending the outpatient and inpatient services of the Department of Pediatrics were screened for eligibility. A total of 1,000 children were enrolled. Inclusion criteria comprised age ≤10 years and a confirmed diagnosis of pulmonary hypertension (PH) established either by right heart catheterization (RHC) based on hemodynamic parameters or by echocardiographic assessment with supportive clinical features, as per multidisciplinary PH team consensus.

Children with pulmonary hypertension secondary to other etiologies (e.g., congenital heart disease, lung disease, left heart disease, or systemic disorders) were excluded. Only those with idiopathic pulmonary arterial hypertension (IPAH) were included in the final analysis cohort.

Data Collection

For each eligible participant, detailed demographic and clinical data were recorded, including age, sex, presenting complaints, and family history. Comprehensive physical examination was carried out with emphasis on cardiopulmonary evaluation.

The clinical symptoms and signs were specifically documented.

Diagnostic Evaluation

All enrolled children underwent non-invasive echocardiography to evaluate right heart structure, pulmonary artery pressure, and ventricular function. In selected cases, right heart catheterization was performed when clinically indicated, applying the following hemodynamic criteria for PH diagnosis:

- Mean pulmonary arterial pressure (mPAP) >20 mmHg at rest
- Pulmonary arterial wedge pressure (PAWP) ≤15 mmHg
- Pulmonary vascular resistance index (PVRi) >3 Wood Units·m²

Children meeting the above criteria without an identifiable secondary cause were classified as having IPAH.

Outcome Measures

The primary outcome was the incidence of idiopathic pulmonary hypertension among the study population. Secondary outcomes included the distribution of IPAH cases by age group and gender, and the frequency of clinical symptoms and signs.

Statistical Analysis

Data were entered into Microsoft Excel and analyzed using descriptive statistical methods. Continuous variables were expressed as mean ± standard deviation (SD), and categorical variables were summarized as frequencies and percentages. Results were presented in tabular form for clarity.

Table 1: Prevalence of Idiopathic pulmonary hypertension according to gender

Gender	Frequency	Percentage
Male	30	60.00
Female	20	40.00
Total	50	100.00

Table 2: Prevalence of Idiopathic pulmonary hypertension according to age

Age groups (yrs)	Frequency	Percentage
1-5	40	80.00
5-10	10	20.00
Total	50	100.00

Table 3: Clinical Symptoms of Idiopathic pulmonary hypertension

Clinical Symptoms	N(%)
Functional dyspnea	
Yes	30(60%)
No	20(40%)
Shortness of breath	
Yes	40(80%)
No	10(2%)
Syncope	
Yes	20(40%)
No	30(60%)

Cyanosis	
Yes	10(20%)
No	40(80%)
Palpitations	
Yes	0(0%)
No	50(100%)
Heart murmurs	
Yes	10(20%)
No	40(80%)
Chest pain	
Yes	40(80%)
No	10(20%)
Edema	
Yes	30(60%)
No	20(40%)
Hepatomegaly	
Yes	20(40%)
No	30(60%)
Ascites	
Yes	20(40%)
No	30(60%)

RESULTS

A total of 1,000 children were evaluated during the six-month study period. Of these, 50 children (5%) were diagnosed with idiopathic pulmonary arterial hypertension (IPAH) based on clinical and echocardiographic assessment.

Demographic Characteristics

Among the 50 affected children, 30 were male (60%) and 20 were female (40%), giving a male-to-female ratio of 1.5:1 (Table 1). The majority of cases were observed in the 1–5 year age group (n = 40, 80%), whereas only 10 children (20%) were between 5 and 10 years of age (Table 2).

Clinical Presentation

The clinical spectrum of IPAH in the cohort is summarized in Table 3. The most frequently reported symptom was shortness of breath (n = 40, 80%), followed by chest pain (n = 40, 80%). Functional dyspnea and peripheral edema were observed in 30 children each (60%). Syncope, hepatomegaly, and ascites were each present in 20 children (40%). Less common findings included cyanosis (n = 10, 20%) and heart murmurs (n = 10, 20%). Notably, no child reported palpitations.

Summary of Findings

Overall, IPAH accounted for 5% of all children assessed during the study period. The condition was more prevalent in males and predominantly affected children in the early age group (1–5 years). Respiratory complaints such as dyspnea and chest pain were the leading symptoms, while systemic manifestations like edema and hepatomegaly were also frequent.

DISCUSSION

Idiopathic pulmonary arterial hypertension (IPAH) represents one of the most significant subtypes of pulmonary arterial hypertension. In earlier studies, IPAH was reported as the most frequent type of pulmonary hypertension, accounting for approximately 69.8% of cases, followed by pulmonary hypertension due to left heart disease (16.9%) and valvular heart disease (10.7%).[7] However, in children, IPAH remains a rare entity, with an estimated incidence of 1–2 cases per million per year and a prevalence of less than 10 per million.[8]

In the present institutional study of 1,000 children, IPAH was diagnosed in 50 cases, yielding an incidence of 5%. A clear male predominance (60%) was observed, and the majority of cases (80%) occurred in children aged 1–5 years. These findings suggest that IPAH tends to present in early childhood with a slight male bias.

Our results are partially consistent with prior cohort data. Moledina et al. reported an incidence of 0.48 cases per million children per year and a prevalence of 2.1 cases per million. In their cohort, 31% of patients presented with syncope, while edema was infrequently noted. Importantly, survival rates at 1, 3, and 5 years were 89%, 84%, and 75%, respectively, with transplant-free survival declining to 57% at 5 years. Prognostic factors included poor functional class at presentation and impaired growth parameters.[9]

Similarly, Yang et al. analyzed a large dataset and found that pulmonary arterial hypertension accounted for 48% of cases, whereas pulmonary hypertension secondary to lung disease was present in 32.3%.[10] Interestingly, 16.9% of patients demonstrated overlapping features from multiple etiological groups. The annual incidence of childhood pulmonary hypertension was estimated at 3.5 per million, with the highest rates observed in infants with lung disease (15.0 per million per year). Long-term outcomes revealed a transplant-free survival of 86.7% at 1 year, falling to 68.6% at 10 years, with pulmonary hypertension due to left heart disease showing the poorest prognosis.[10]

In our study, the predominant symptoms were shortness of breath (80%) and chest pain (80%), followed by functional dyspnea (60%) and peripheral edema (60%). Syncope, hepatomegaly, and ascites were reported in 40% of cases, while cyanosis and

heart murmurs were observed in 20%. Notably, palpitations were absent in all children. These findings are broadly in line with existing literature, where dyspnea has been reported in over 90% of pediatric PH cases.[11] Barst et al[12] and Charalampopoulos et al[13] also identified dyspnea, shortness of breath, syncope, cyanosis, and edema as among the most common clinical manifestations of pediatric PH. Taken together, these findings reinforce that although IPAH is relatively rare in children, it is associated with significant morbidity. Early recognition of hallmark symptoms, particularly dyspnea and chest pain, is critical for timely diagnosis and initiation of therapy.

CONCLUSION

In this institutional study, the overall incidence of idiopathic pulmonary arterial hypertension (IPAH) among children was found to be 5%, with the majority of cases occurring in the 1–5 year age group and showing a slight male predominance. Dyspnea, chest pain, and systemic features such as edema and hepatomegaly were the most frequent clinical manifestations.

These findings highlight the need for heightened clinical vigilance in pediatric populations presenting with unexplained dyspnea, chest discomfort, or signs of right heart strain. Routine use of non-invasive echocardiographic screening in such cases may facilitate earlier identification of IPAH, thereby enabling timely initiation of therapy and potentially improving long-term outcomes. Furthermore, strengthening awareness among pediatricians regarding the subtle and variable presentations of IPAH is crucial for reducing diagnostic delays and optimizing patient survival.

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