The Anesthetic Management of Children with Pulmonary Hypertension in the Cardiac Catheterization Laboratory

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KEYWORDS

- Pulmonary hypertension • Cardiac catheterization laboratory • Anesthesia
- Children

KEY POINTS

- A new classification of pediatric pulmonary arterial hypertension (PAH) has been developed that incorporates abnormalities of lung growth and development as well as syndromes frequently contributing to PAH.
- Children with PAH will require cardiac catheterization to establish the diagnosis and monitor the response to therapy.
- Children receiving general anesthesia for cardiac catheterization are at significantly increased risk of perioperative complications such as a pulmonary hypertensive crisis.
- There is no one ideal anesthetic agent for children with PAH, and it is essential to understand the different hemodynamic effects of anesthetic agents and adopt a balanced anesthetic technique for children with PAH.

INTRODUCTION

Pulmonary hypertension has many different causes, which all share the final common pathway of elevated pulmonary arterial pressure (PAP). Pulmonary arterial hypertension (PAH) is due to abnormalities in the pulmonary arterial vasculature. Pulmonary venous hypertension is a result of left-sided heart disease, for example, pulmonary vein stenosis or left-side valvar heart disease. The treatments of PAH and pulmonary venous hypertension are different, so the distinction of one from the other is of obvious clinical importance.[1] This article focuses on PAH in children. PAH is a life-threatening disease which, if undiagnosed, will eventually culminate in irreversible elevation of

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pulmonary vascular resistance (PVR), leading to right ventricular failure and death.[2] Unfortunately, delays in diagnosis and treatment are not uncommon because of nonspecific presenting symptoms, especially in young children, and the low incidence of the disease.[3] The estimated prevalence in adults is 15 to 50 cases per 1 million, whereas in children it is less than 10 cases per 1 million.[4,5]

Children with suspected PAH require invasive hemodynamic assessment in the cardiac catheterization laboratory to confirm the diagnosis of PAH and determine future therapy. To tolerate the procedure, most of these children will need general anesthesia provided by an anesthesiologist. It is essential, therefore, that the providing anesthesiologist understands the pathophysiology of PAH, which measurements are made in the catheterization laboratory, how anesthetic medications may affect these measurements, and how to manage a pulmonary hypertensive crisis.[6–9] Children with PAH, especially those with a new diagnosis who are not yet on any treatment, are at increased risk of complications under anesthesia in the cardiac catheterization laboratory. The cardiologist performing the procedure, the pediatric anesthesiologist, and the catheterization laboratory support staff must effectively communicate to provide safe perioperative care.

DEFINITION AND CLASSIFICATION

In normal, healthy individuals the mean pulmonary artery pressure (mPAP) at rest is around 15 mm Hg, and is independent of age, ethnicity, and gender. During exercise, mPAP increases and is dependent on the level of exertion and age. During mild exercise, mPAP is 20 $\pm$ 5 mm Hg in subjects younger than 50 years compared with 30 $\pm$ 5 mm Hg in subjects older 50, which makes it difficult to define normal mPAP during exercise; hence, the definition of PAH uses mPAP at rest.[10] PAH is defined as mPAP greater than 25 mm Hg at rest, with a normal pulmonary capillary wedge pressure ($\leq$15 mm Hg) and increased pulmonary vascular resistance index (PVRI) greater than 3 Wood units per m^2.[11] The normal pulmonary capillary wedge pressure excludes patients with pulmonary venous hypertension from left-sided heart disease. In patients with suspected PAH, the initial investigation is usually a transthoracic echocardiogram that can estimate the mPAP and diagnose any congenital cardiac lesions that may be contributing to the PAH. Echocardiography may support the diagnosis of PAH with qualitative images of elevated right ventricular pressure, such as right ventricular hypertrophy and septal-wall flattening. Quantitative information may be obtained on echocardiography if there is tricuspid regurgitation during systole. In this case, the modified Bernoulli equation may be applied to estimate mPAP, with a tricuspid regurgitant velocity of greater than 2.8 m/s being highly indicative of PAH (**Box 1**).[10,12]

Transthoracic echocardiography is an attractive method to monitor children with PAH, and possibly enable the cardiologist to lengthen the interval between cardiac catheterizations that the child will require to monitor ongoing therapy. There are many echocardiographic techniques in the research and validation phase. One technique is to monitor the right ventricular systolic to diastolic duration ratio, whereby an increase has been shown to be associated with worse right ventricular function, exercise capability, and survival.[13] Another is to measure the degree of tricuspid annular plane systolic excursion (TAPSE), which has been shown to reflect right ventricular function and prognosis in PAH.[14,15]

In the absence of shunts, the pulmonary and systemic circulations receive the same amount of blood flow per minute. PVR beyond the newborn period is more than 10-fold lower than resistance in the systemic circulation, and the pressure in the venous bed draining the pulmonary arteries (pulmonary veins, left atrium) accounts