

Functional classification of pulmonary hypertension in children: Report from the PVRI pediatric taskforce, Panama 2011

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ABSTRACT

The members of the Pediatric Task Force of the Pulmonary Vascular Research Institute (PVRI) were aware of the need to develop a functional classification of pulmonary hypertension in children. The proposed classification follows the same pattern and uses the same criteria as the Dana Point pulmonary hypertension specific classification for adults. Modifications were necessary for children, since age, physical growth and maturation influences the way in which the functional effects of a disease are expressed. It is essential to encapsulate a child's clinical status, to make it possible to review progress with time as he/she grows up, as consistently and as objectively as possible. Particularly in younger children we sought to include objective indicators such as thriving, need for supplemental feeds and the record of school or nursery attendance. This helps monitor the clinical course of events and response to treatment over the years. It also facilitates the development of treatment algorithms for children. We present a consensus paper on a functional classification system for children with pulmonary hypertension, discussed at the Annual Meeting of the PVRI in Panama City, February 2011.

Key Words: pulmonary hypertension, children, functional class

INTRODUCTION

The members of the Pediatric Task Force of the Pulmonary Vascular Research Institute (PVRI) were very aware of the need to develop a functional classification system for children with pulmonary hypertension and discussed the problem at the Annual Meeting of the PVRI held in Panama City in February 2011. We now present the consensus document resulting from our deliberations. We expect that it will be modified and improved in the years to come.

BACKGROUND

Clinical classifications of functional status are created

primarily so that physicians can apply a common language to describe the functional impact of the same underlying disease on individual patients. Classifications should facilitate communication between physicians, all those involved in caring for the patient and the funding agencies providing financial support. The design of clinical trials is heavily dependant on an accepted, standardized means of describing the efficacy or otherwise of the treatment being evaluated. Studying the natural history of a disease, both treated and untreated, is dependant on a universally accepted means of describing functional status. Most importantly, a good classification should encapsulate

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the clinical status of a patient and so make it possible to review the patient's progress with time and their response to treatment.

Classifications of functional status are familiar to cardiologists caring for adult patients. The New York Heart Association (NYHA), the classification most widely used by adult cardiologists since 1964 describes the functional impact of heart failure and places patients with a similar degree of limitation and similar symptoms into one of four functional classes, Class IV being the most severely disabled (Table 1).^[1] The Functional Classification of Pulmonary Hypertension in adults is based on the NYHA classification (Table 2) and was published in 1998 as a consensus document of the WHO Symposium held in Evian in that year.^[2]

Maintaining the best possible quality of life is crucial in any chronic disease. In adults, The Minnesota Living with Heart Failure (MLHF) questionnaire has been widely used in both hospital and primary care since it was designed in 1984, and has good reliability and validity.^[3-5] This questionnaire can be useful in assessing patients with pulmonary hypertension,^[6,7] as is the Short Form Health Survey 36 (SF-36).^[8,9] The MLHF has been considered a significant predictor of outcome.^[10] More recently, a disease specific questionnaire, the Cambridge Pulmonary Hypertension Outcome Review Utility Index (CAMPHOR) has been developed, primarily for cost-utility analysis but can also be useful in clinical studies.^[11,12]

There is no disease specific classification to assess

functional status in children with pulmonary hypertension. Nor is there a generally accepted functional classification for children with heart disease. Measures of generic health status have been developed for children, principally the Children and Youth Version of the International Classification of Functioning, Disability and Health published in 2007.^[13,14] This classification assesses body structure and function, level of activity and social participation. It was designed primarily to assess children with neuromotor disabilities. Disease specific functional classifications have been developed for use in children with other conditions such as cystic fibrosis, rheumatoid arthritis and juvenile idiopathic arthritis.^[15-18]

Quality of life is particularly difficult to assess in children. A Short Form Health Survey has been designed for children, SF-10^[19] and can be used in children with pulmonary hypertension, but it is not disease specific. Nor does it help assess children less than 5 five years of age.

DESIGNING THE PROPOSED FUNCTIONAL CLASSIFICATION OF PULMONARY HYPERTENSION IN CHILDREN

The proposed Functional Classification of Pulmonary Hypertension in Children is based on the Evian pulmonary hypertension specific classification used in adults. It is not designed to assess quality of life and neither parent nor child makes a personal, direct contribution to the assessment. Particularly in children however, assessing

Table 1: NYHA classification of functional status

NYHA Class	Symptoms
I	Patients with heart disease but without limitation of physical activity
II	Patients with heart disease resulting in slight limitation of physical activity Comfortable at rest
III	Patients with heart disease resulting in marked limitation of physical activity. Comfortable at rest. Less than ordinary activity causes fatigue, dyspnoea, palpitations, anginal pain
IV	Patients with heart disease resulting in inability to carry out any physical activity without discomfort. Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort increases

Table 2: WHO classification of pulmonary hypertension in adults

Class	Symptoms
I	Patients with pulmonary hypertension but without limitation of physical activity. Ordinary physical activity does not cause undue dyspnoea, fatigue chest pain or near syncope
II	Patients with pulmonary hypertension resulting in slight limitation of physical activity. Comfortable at rest. Ordinary physical activity causes undue dyspnoea, fatigue, chest pain or near syncope
III	Patients with pulmonary hypertension resulting in marked limitation of activity. Comfortable at rest. Less than ordinary activity causes dyspnoea or fatigue, chest pain or near syncope
IV	Patients with pulmonary hypertension resulting in inability to carry out any physical activity without symptoms. These patients manifest symptoms of right heart failure. Dyspnoea and/or fatigue may be present even at rest. Discomfort is increased by any physical activity undertaken. Syncope or near syncope can occur

function and activity does inform the physician about the quality of life.

Designing a functional classification is particularly difficult in children because the age, physical growth and maturation achieved influences the way in which the functional effects of a disease are expressed. It is essential therefore to be able to encapsulate a child's clinical status as he/she grows up in a consistent manner which makes it possible to monitor the clinical course of events over the years. In young children it is particularly challenging to distinguish whether any apparent deficiency in expectation can be attributed to the disease or is merely a normal variant in a developmental milestone. This is especially the case if pulmonary hypertension is

complicated by syndromic or chromosomal abnormalities that affect motor and sensory function. Since any parent is naturally loathe to acknowledge that their child is really ill, any clinical classification needs to be as objective as possible.

The proposed Functional Classification of Pulmonary Hypertension in Children follows the same pattern as the adult classification (Tables 3-7). There are four classes of disease severity, Class IV being the most severe. Class III has been subdivided into a) and b). We have tried to ensure that the classification will give a comprehensive summary of the child's clinical condition, while keeping it practical and easy to use. We decided to define functional class in five different age groups (Table 3). Three of the groups

Table 3: Pediatric functional classification for children aged 0–0.5 years

Class	Children with pulmonary hypertension
I	Asymptomatic, growing and developing normally, no limitation of physical activity. Gains head control and increases body tone from 0 to 3 months, then rolls over and has no head lag. Sitting with support
II	Slight limitation of physical activity, unduly dyspnoeic and fatigued. Falling behind physical developmental milestones. Comfortable at rest. Continues to grow along own centiles
IIIa	Marked limitation of physical activity, unduly fatigued. Regression of learned physical activities. Quiet and needs frequent naps. Comfortable at rest. Less than ordinary activity causes undue fatigue or syncope and/or presyncope. Growth compromised. Poor appetite. Requires excessive medical attention
IIIb	Growth severely compromised. Poor appetite. Supplemental feeding. Less than ordinary activity causes undue fatigue or syncope. Plus features of Class IIIa
IV	Unable to carry out any physical activity without undue dyspnoea, fatigue or syncope, not interacting with family. Syncope and/or right heart failure. Plus features of Class III

Table 4: Pediatric functional classification for children aged 0.5–1 years

Class	Children with pulmonary hypertension
I	Asymptomatic, growing along own centiles, no limitation of physical activity. Mobile, sitting, grasping, starting to stand, crawling, playing
II	Slight limitation of physical activity, unduly dyspnoeic and fatigued when playing. Delayed physical development. Comfortable at rest. Continues to grow along own centiles
IIIa	Marked limitation of physical activity. Regression of learned physical activities. Stops crawling. Quiet and needs frequent naps. Hesitant and unadventurous. Comfortable at rest. Less than ordinary activity causes undue fatigue or syncope and/or presyncope. Growth compromised. Poor appetite. Requires excessive medical attention
IIIb	Growth severely compromised. Poor appetite. Supplemental feeding. Less than ordinary activity causes undue fatigue or syncope. Plus features of Class IIIa
IV	Unable to carry out any physical activity without undue dyspnoea, fatigue or syncope, not interacting with family. Syncope and/or right heart failure. Plus features of Class III

Table 5: Pediatric functional classification for children aged 1–2 years

Class	Children with pulmonary hypertension
I	Asymptomatic, growing along own centiles, no limitation of physical activity. Standing, starting to walk/walking, climbing
II	Slight limitation of physical activity, unduly dyspnoeic and fatigued when playing. Delayed physical development. Comfortable at rest. Continues to grow along own centiles
IIIa	Marked limitation of physical activity. Regression of learned physical activities. Reluctant to play. Quiet and needs frequent naps. Hesitant and unadventurous. Comfortable at rest. Less than ordinary activity causes undue dyspnoea, fatigue, or syncope and/or presyncope. Growth compromised. Poor appetite
IIIb	Growth severely compromised. Poor appetite. Supplemental feeding. Less than ordinary activity causes undue fatigue or syncope. Plus features of Class IIIa
IV	Unable to carry out any physical activity without undue dyspnoea, fatigue or syncope, not interacting with family. Syncope and/or right heart failure. Plus features of Class III

deal with children less than two years of age when the most rapid physical development and maturation occurs. Between the ages of two and five years it becomes easier to communicate with the child, while by six years the child is able to describe what they are able to do and how they feel. Children become increasingly articulate with age unless neurodevelopmental problems complicate the assessment. The adult classification is appropriate for those aged 16 years and over. Obviously, the patient is placed in the functional class which best summarises their clinical status and not all the descriptors of a functional class need to be fulfilled in order to place the patient in a particular class.

Bearing in mind that young children cannot tell one

how they feel, we sought to include objective indicators such as whether or not the child is thriving, the need for supplemental feeds and the record of school or nursery attendance. The threshold for supplemental feeding can vary with age and be influenced by comorbidities but the need for supplemental feeding is usually an indication of disease severity. Parents are often reluctant to acknowledge the extent to which their child's schooling is compromised by illness, realising that absence from school reflects how sick their child really is. The days of school missed due to illness provides a measurable yardstick of illness severity. Therefore the classification includes guidelines suggesting an attendance of 75 or 50% or less to encourage them to give a realistic answer.

Table 6: Pediatric functional classification for children aged 2–5 years

Class Children with pulmonary hypertension

I	Asymptomatic, growing normally attending nursery/school regularly, no limitation of physical activity, playing sports with his/her classmates
II	Slight limitation of physical activity, unduly dyspnoeic and fatigued when playing with his/her classmates. Comfortable at rest. Continues to grow along own centiles. Nursery/school attendance 75% normal. No chest pain
IIIa	Marked limitation of physical activity Regression of learned physical activities. Not climbing stairs, reluctant to play with friends. Hesitant and unadventurous. Comfortable at rest. Less than ordinary activity (dressing) causes undue dyspnoea, fatigue, syncope and/or presyncope or chest pain. Nursery/schooling compromised <50% normal attendance
IIIb	Unable to attend nursery/school, but mobile at home. Wheelchair needed outside home. Growth compromised. Poor appetite. Supplemental feeding. Less than ordinary activity causes undue fatigue, syncope or chest pain. Plus features of Class IIIa)
IV	Unable to carry out any physical activity without undue dyspnoea, fatigue, syncope or chest pain, unable to attend school, wheelchair dependant, not interacting with friends. Syncope and/or right heart failure. Plus features of Class III

Table 7: Pediatric functional classification for children aged 5–16 years

Class Children with pulmonary hypertension

I	Asymptomatic, growing along own centiles, attending school regularly, no limitation of physical activity, playing sports with his/her classmates
II	Slight limitation of physical activity, unduly dyspnoeic and fatigued when playing when playing with his/her classmates. Comfortable at rest. Continues to grow along own centiles. School attendance 75% normal. No chest pain
IIIa	Marked limitation of physical activity. No attempt at sports. Comfortable at rest. Less than ordinary activity causes undue dyspnoea, fatigue, syncope or chest pain. Schooling compromised, <50% normal attendance
IIIb	Unable to attend school, but mobile at home and interacting with friends. Wheelchair needed outside the home. Growth compromised. Poor appetite. Supplemental feeding. Less than ordinary activity (dressing) causes undue dyspnoea, fatigue, syncope and/or presyncope or chest pain. Plus features of Class IIIa)
IV	Unable to carry out any physical activity without undue dyspnoea, fatigue, syncope or chest pain, unable to attend school, wheelchair dependant, not interacting with friends. Syncope and/or right heart failure. Plus features of Class III

Table 8: What to expect of children in Functional Class I

Age	Children with pulmonary hypertension, asymptomatic, growing and developing normally, no limitation of physical activity
0-0.5 years	Gains head control and increases body tone from 0 to 3 months, then rolls over and has no head lag. Sitting with support
0.5-1 year	Mobile, sitting, grasping, starting to stand, crawling, playing
1-2 years	Standing, starting to walk/walking, climbing
2-5 years	Attending nursery/school regularly, playing games with his/her peers
6-15 years	Children attending school regularly, playing games with his/her peers

DESCRIPTORS OF FUNCTIONAL CLASS

Functional Class I

This is defined, at all ages, as children with pulmonary hypertension who are asymptomatic, growing and developing normally and have no limitation of physical activity. Table 8 outlines the most important features seen in normal children in the five age groups used in this classification and which we expect to find in children categorised as functional Class I. The emphasis on motor development in the first two years of life gradually shifts to the child's ability to interact with his/ her peers, participate in sporting activities and go to nursery and then school.

Functional Class II

At all ages children with pulmonary hypertension are categorised as Functional Class II when they have only a slight limitation of physical activity due to fatigue and or dyspnoea but are comfortable at rest. Dyspnoea can interrupt feeding in young children. At this stage of their disease most children do not experience syncope or presyncope but some children can do so while still having a good exercise tolerance. During the first six months of life they fall behind their developmental milestones but continue to grow along their own centiles. Infants and young children are readily fatigued and dyspnoeic when playing. Beyond two years of age, it is important to assess attendance at school or nursery, which should be at least 75% that of healthy children.

Functional Class IIIa

Functional Class IIIa is characterised by marked limitation of physical activity. In addition to failing to achieve their developmental milestones, children between the ages of six months and two years may show regression of newly learnt activities. Inactivity is noticeable, the child being quiet and taking frequent naps. Less than ordinary activity such as dressing is tiring and can cause dyspnoea. Children frequently experience syncope and /or presyncope. Older children can become withdrawn and less confident, choosing to spend time with their families rather than their friends. Growth is compromised and appetite is poor. Nursery/school attendance is less than 50% of normal. The parents frequently say that the child has required excessive medical attention.

Functional Class IIIb

In addition to the features characteristic of Class IIIa, children in Class IIIb often require supplemental feeding by nasogastric tube or gastrostomy. Older children can no longer go to school and although mobile at home they need a wheelchair when venturing out of doors.

Functional Class IV

Children in Class IV are severely compromised and unable to carry out any physical activity without fatigue and /or dyspnoea. They are frequently syncopal, may complain of chest pain and often become quiet and withdrawn. Signs of right heart failure are frequently present, particularly in teenagers.

LIMITATIONS OF THE PROPOSED FUNCTIONAL CLASSIFICATION FOR CHILDREN

The growth and development of healthy children varies considerably and children with pulmonary hypertension will be subject to the same innate variability, increasing the difficulty facing the physician trying to assess the functional impact of disease. In addition, many children with pulmonary hypertension have multisystem disorders that may impact heavily on functional capacity independently of their pulmonary vascular disease. We have tried to ensure that the features characteristic of each functional class are objective but the opinion of the parents can influence judgement, particularly in a first child of young parents without extended family support who maybe uncertain about how their child differs from the normal. Some children are more stoical than others. Some physicians will recommend supplemental feeding more readily than others. Comorbidity further complicates assessment, primarily in children with neurodevelopmental handicap who may be syndromal. Despite all these and other caveats the Pediatric Task Force of the Pulmonary Vascular Research Institute believes that the proposed classification will facilitate the management of children with pulmonary hypertension.

CONCLUSIONS

The proposed Functional Classification of Pulmonary Hypertension in Children follows the same pattern and uses the same criteria as the adult classification, modified appropriately for children of all ages. The pediatric classification should therefore be understood readily by adult physicians during transition from pediatric to adult medical services, by all those caring for children with pulmonary hypertension and by those carrying out clinical research and designing clinical trials. It should facilitate the development of treatment algorithms for children, as has the adult classification in older patients with pulmonary hypertension.^[20] This is a consensus document and we hope and expect that it will be improved upon with experience.

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