Recommendations from the Association for European Paediatric and Congenital Cardiology for training in pulmonary hypertension*

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*On behalf of the Association for European Paediatric and Congenital Cardiology working group "Pulmonary Hypertension, Heart Failure and Transplantation".

Abstract

Pulmonary hypertension is a complex and progressive condition that is either idiopathic or heritable, or associated with one or multiple health conditions, with or without congenital or acquired cardiovascular disease. Recent developments have tremendously increased the armamentarium of diagnostic and therapeutic approaches in children and young adults with pulmonary hypertension that is still associated with a high morbidity and mortality. These modalities include non-invasive imaging, pharmacotherapy, interventional and surgical procedures, and supportive measures. The optimal, tailored diagnostic and therapeutic strategies for pulmonary hypertension in the young are rapidly evolving but still face enormous challenges: Healthcare providers need to take the patient’s age, development, disease state, and family concerns into account when initiating advanced diagnostics and treatment. Therefore, there is a need for guidance on core and advanced medical training in paediatric pulmonary hypertension. The Association for European Paediatric and Congenital Cardiology working group “pulmonary hypertension, heart failure and transplantation” has produced this document as an expert consensus statement; however, all recommendations must be considered and applied in the context of the local and national infrastructure and legal regulations.

Background

Pulmonary hypertension is a complex and progressive condition that is either idiopathic or heritable, or associated with one or multiple health conditions, with or without congenital or acquired cardiovascular disease. Recent developments have tremendously increased the armamentarium of diagnostic and therapeutic approaches in children and young adults with pulmonary hypertension that is still associated with a high morbidity and mortality. These modalities include non-invasive imaging, pharmacotherapy, interventional and surgical procedures, and supportive measures. However, due to a lack of robust clinical trial data in children and adolescents, most of these new agents (drugs and devices) have not yet been approved for paediatric use by the regulatory authorities. Thus, many medications are used off-label by paediatric pulmonary hypertension specialists. Recently, consensus statements and according recommendations that specifically address the care of children with pulmonary hypertension have been developed (the majority with level of evidence B or C).

Specialised care for paediatric and congenital cardiology patients with pulmonary hypertension is rapidly evolving and should follow a comprehensive multi-disciplinary approach with a number of complementary medical, surgical, and supportive avenues, which may include cardiac catheterisation, non-invasive imaging (echocardiography and MRI), radiology, blood tests, pulmonary function, pharmacotherapy, and lung transplantation. Furthermore, the pivotal importance of psycho-social support and care for patients and their families needs to
be acknowledged as pulmonary hypertension represents both a critical illness and a chronic, progressive disease that currently is without a cure.

The aforementioned challenges, recent developments, and specifics of pulmonary hypertension in paediatric and congenital cardiology underline the need for guidance for core and advanced medical training in this specific area. Thus, the Association for European Paediatric and Congenital Cardiology (AEPC) working group “pulmonary hypertension, heart failure and transplantation” has produced this document as an expert consensus statement on clinical training in paediatric pulmonary hypertension; however, all recommendations must be considered and applied in the context of the local and national infrastructure structure and legal regulations.

“Few areas in clinical medicine are as challenging – intellectually, technically, and emotionally – as caring for a child with serious and often life-threatening pulmonary vascular disease.” Lewis Rubin, MD (University of California San Diego)

General considerations

The development of training requirements is the responsibility of the Educational Committee and the Council of the AEPC, in collaboration with the working groups. The recommendations presented in this document are based on a consensus reached by the authors. Face-to-face meetings, telephone conferences, and written communication among the members of the writing group were used during the development and writing process of this consensus statement (2017–2019).

As a result, we outline here the recommendations of the AEPC for training in paediatric pulmonary hypertension. Many of these recommendations are defined “qualitatively” and not necessarily “quantitatively”. Trainees in paediatric cardiology should be exposed to all aspects of general paediatric and congenital cardiology from foetal life to adolescence and adulthood. All paediatric cardiologists should achieve the basic training standards (core level) with those wishing to specialise in pulmonary hypertension achieving more advanced training (advanced level). Fellows seeking advanced training in pulmonary hypertension must have completed core training in paediatric and congenital cardiology, which usually encompasses at least 3 years of training, as outlined by the AEPC’s Educational Committee (available on the website www.aepc.org). The specific requirements for advanced training in paediatric pulmonary hypertension will be discussed in a separate section later in this guideline. Of note, paediatric pulmonologists and neonatologists frequently take care of children with pulmonary hypertension, especially those in group 3 or group 5 pulmonary hypertension (as defined by the World Symposium on Pulmonary Hypertension in 2018). Addressing the specific training needs and knowledge gaps of both pulmonologists and neonatologists is beyond the scope of this training guideline. However, the best clinical results for children with pulmonary hypertension are reached through a collegial, interdisciplinary team effort which should be supervised by a paediatric cardiologist with expertise in pulmonary hypertension.

Programme and training centre requirements

Pulmonary hypertension is a well-recognised complication of CHD but is also encountered as idiopathic/heritable disease or in association with other systemic diseases, leading to cardiology referral by other paediatric subspecialty departments (e.g. neonatology, pulmonology, rheumatology, haematology). Therefore, it is imperative that all paediatric cardiology training programmes cover the core training requirements as outlined next.

Advanced training for cardiologists who wish to specialise in pulmonary hypertension requires direct experience of additional diagnostic techniques and treatments. It is recognised that this may pose a challenge given the rare nature of pulmonary hypertension. Training programmes may wish to incorporate rotations to centres with high volume or particular expertise with a specific treatment or diagnostic modality or disease entity. Training may also be supported by attendance of conferences and workshops.

Core training institutions should be high-volume centres that have both a core fellowship in paediatric/congenital cardiology and an established pulmonary hypertension programme. Ideally the centre would assess more than 10–15 newly diagnosed children with pulmonary hypertension (any aetiology) per year and provide clinical follow-up for 30–50 children of a variety of disease entities and disease stages, including (but not limited to) idiopathic pulmonary arterial hypertension, heritable pulmonary arterial hypertension, CHD associated pulmonary hypertension, pulmonary hypertension associated with left heart or lung diseases (e.g. bronchopulmonary dysplasia, parenchymal/interstitial lung disease), and pulmonary hypertension associated with systemic diseases (e.g. connective tissue disease and haematological disorders).

The centre(s) dedicated to advanced training should have a well-developed pulmonary hypertension service involving a number of integrated multi-disciplinary teams and specialists, containing but not limited to: paediatric/congenital cardiologists, paediatric/congenital cardiac surgery, paediatric pulmonologist, imaging specialists (echocardiography and MRI), pharmacology, genetics, psychosocial/family education and support. Furthermore, a paediatric cardiologist seeking sub-specialty training in pulmonary hypertension should have exposure to patients in both an ambulatory and an in-hospital setting, which should also include access to critical care facilities (e.g. cardiac and/or paediatric intensive care, neonatology).

We recommend the centre(s) engaged in advanced training should have regular multi-disciplinary meetings specifically discussing the management of pulmonary hypertension patients. Furthermore, a dedicated centre for advanced training in paediatric pulmonary hypertension should have diagnostic, treatment, and follow-up protocols in place. In addition, the core training centres should be actively involved in research relevant to the field (basic, clinical, and/or translational).

Principles of teaching, supervision, and assessment

Clinical care and education should be supervised by experts specifically assigned to these tasks (e.g. programme and/or fellowship director). Centres performing generalised and specialised work in paediatric and congenital cardiology should be committed to delivering post-graduate training. Core training must include sufficient clinical exposure and didactics to achieve competency in the evaluation and management of children with pulmonary hypertension. The AEPC provides and/or endorses basic teaching courses and practical workshops to supplement the training process.

Successful clinical training is based on a variety of teaching and learning methods, which include (but are not limited to) experiential learning, formal teaching, and independent self-directed learning (Table 1). Constant supervision plays a pivotal role in the teaching process. Trainees should have a named clinical/educational supervisor, responsible for overseeing their training/education and logbook that should be part of the clinical specialty team.
The appointed supervisor should be an experienced clinician (e.g. pulmonary hypertension fellowship director) who holds regular consultations between trainee and supervisor to discuss patients, but also the progress of the training process. Opportunities for feedback to trainees about their performance will arise from the supervisor, colleagues, and feedback from an annual assessment review by a designated panel. Together with the trainee, the supervisor and the fellowship oversight committee share the responsibility for integrating non-clinical education (e.g. research activity, conference attendance, teaching delivered by the trainee) into the individual curriculum of each trainee. A constant monitoring and assessment is imperative to the success of every training process. Thus, besides structured supervision and feedback, fulfillment of all educational and curricular requirements should be documented in a structured logbook, preferably electronically, which will be evaluated and accepted by the responsible supervisor (paediatric cardiology fellowship director or the division chief of pulmonary hypertension). A more detailed discussion on education and training in paediatric cardiology can be found in previously published documents.10–12

Training curriculum

The recommendations outlined next should be regarded as the minimum requirements in pulmonary hypertension to become a paediatric and congenital cardiologist (basic), with or without additional comprehensive training in pulmonary hypertension (advanced). Table 1 provides a summary of paediatric and congenital cardiology training in pulmonary hypertension.

### Core (basic) training in pulmonary hypertension

The basic level is recommended for all paediatric cardiology trainees and should be available at all centres/networks with a general training programme in paediatric/congenital cardiology (core fellowship, usually 3 years). The goal of such training is to equip the trainee with knowledge and skills to:

- Detect and diagnose pulmonary hypertension in patients presenting with or without symptoms, especially in those from high-risk populations.4
- Perform an initial evaluation of the child with pulmonary hypertension in the outpatient setting, identifying likely contributory factors/risk factors.
- Initiate appropriate supportive care.
- Perform the initial evaluation and stabilisation of the haemodynamically compromised patient with pulmonary hypertension.
- Understand the indications and appropriate timing of referral to a dedicated specialist in paediatric or adult pulmonary hypertension for advanced care.

### Knowledge

- Basic knowledge on pathophysiology of perinatal transition, lung development, and normal pulmonary vascular physiology.
- Recognition of the signs and symptoms of pulmonary hypertension in children, ability to make a differential diagnosis and know when and how to initiate treatment.
- Basic understanding of the pathophysiology and aetiologies, subgroups, classification and natural history of pulmonary hypertension.
- Familiarity with a variety of diagnostic tools to establish the diagnosis and confirm the aetiology of pulmonary hypertension such as non-invasive imaging (echocardiogram and MRI), radiology, blood tests, lung function, exercise testing.
- Evaluation and interpretation of diagnostic cardiac catheterisation to confirm the diagnosis of pulmonary hypertension, categorise the sub-type, and quantify the severity and vasoreactivity. Cardiac catheterisation should be performed by a physician, trained and experienced in the invasive diagnostics of paediatric pulmonary hypertension/pulmonary arterial hypertension.5
- Recognition and quantification of elevated pulmonary vascular resistance in patients with CHD.

<table>
<thead>
<tr>
<th>Table 1. Summary of paediatric and congenital cardiology training in pulmonary hypertension</th>
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<tr>
<td><strong>Core Training in Pulmonary Hypertension</strong></td>
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<tr>
<td><strong>Training Programme Curriculum</strong></td>
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<tr>
<td>- Medical knowledge, skills, attitudes, and understanding of PH/PAH (see main text)</td>
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<td>- Core knowledge base for PH/PAH in children and ACHD (see main text)</td>
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<td><strong>Teaching and Learning Methods</strong></td>
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<td>- Learning with peers</td>
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<td>- Paediatric cardiology on-call</td>
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<td>- Consultant-led ward rounds</td>
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<td>- MDT meetings</td>
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<td>- Formal post-graduate teaching</td>
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<td>- Independent self-directed learning</td>
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<td><strong>Assessment</strong></td>
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<td>- Work place-based clinical assessment, case discussion, patient surveys</td>
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<td>- Supervision, regular and structured feedback, including MSF</td>
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<td>- Record of assessment, progress logbook, portfolio of educational activities</td>
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<td><strong>Advanced Training in Pulmonary Hypertension</strong></td>
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<td>- 2 years in recognised centre (12 months in PH and another 12 months in a complementary subspecialty such as heart failure and transplantation, interventional cardiology, non-invasive imaging, intensive care, adults with CHD)</td>
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<td>- Well-developed clinical PH service</td>
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<td>- Adequate volume of outpatient PH patient visits</td>
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<td>- Experience in an ACHD outpatient clinic</td>
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<tr>
<td>- Adequate volume of inpatient PH patients, including ICU exposure (neonatal, pediatric and/or cardiac, ECMO, intravenous PCA therapy)</td>
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<td>- Elective or regular training in a HTx/LuTx center</td>
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<td>- Regular PH and (optional) HTx/LuTx MDT meetings</td>
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<td>- Exposure to surgery, for example, HTx, LuTx, VAD, ECMO cannulation, Potts shunt</td>
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<td>- Participation in research and attendance/presentation at international meetings</td>
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ACHD = adult congenital heart disease; ECMO = extracorporeal membrane oxygenation; HTx = heart transplantation; ICU = intensive care unit; LuTx = lung transplantation; MDT = multi-disciplinary team; MSF = multi-source feedback; PAH = pulmonary arterial hypertension; PCA = prostacyclin analogues; PH = pulmonary hypertension; VAD = ventricular assist device.
• Understanding the principles of medical management of pulmonary hypertension (acute, chronic, and decompensated).
• Familiarity with diuretics, inotropes, anticoagulation and commonly used pulmonary vasodilators, and age-dependent variations in dosing, both in the intensive care setting and in the outpatient setting.
• Understanding of the place of additional therapies in pulmonary hypertension management, including interventional/surgical procedures such as creation of an interatrial communication and reverse Potts procedure (surgical/interventional: duct stenting or de novo).
• Knowledge on the indications and contraindications for repair of CHD in presence of elevated pulmonary vascular resistance.

Skills
• History and examination for presence and aetiology of pulmonary hypertension.
• Ability to use the Fick principle and thermodilution to calculate cardiac output and shunt volume.
• Echocardiographic assessment of pulmonary hypertension including diagnosis, physiology, shunts, presence of relevant CHD, assessment of afterload and ventricular function.
• Recognise deterioration and know how to escalate (e.g. need to refer to specialist to escalate therapy or for transplantation assessment).

Attitude
• Ability to communicate sensitively and with compassion.
• Ability to break bad news.
• Recognition of importance of multi-disciplinary team (e.g. nurses and psychology) in care for children with pulmonary hypertension.
• Recognise the difference between core (basic) training and advanced training in paediatric pulmonary hypertension, including associated referral indications.

Advanced training in pulmonary hypertension
Those wishing to specialise in this field will require an additional period of training in an accredited specialist centre in order to achieve the curriculum competencies, obtaining comprehensive experience in paediatric pulmonary hypertension (24 months). The advanced trainee would be expected to spend at least half of his or her advanced training (equivalent to 12 months) involved in the direct care of pulmonary hypertension patients (inpatient and outpatient), including clinics, echocardiography, multi-disciplinary meetings, cardiac catheterisation, MRI or cardiopulmonary exercise testing, and analysis of data from these sessions. Time spent in adult pulmonary hypertension is actively encouraged; however, at least the equivalent of 9 months (over the 24-month period) should be spent in paediatric pulmonary hypertension training.

The remaining training period (12 months) may – and probably should – be spent in one complimentary paediatric/congenital cardiac subspecialty, according to those that are available at the institution and carry sufficient annual patient volume, for example:

- heart failure and transplantation,
- interventional cardiology,
- non-invasive imaging,
- intensive care, and
- adults with CHD.

Knowledge
• Comprehensive knowledge of pathobiology and genetics of different groups of pulmonary hypertension.
• Understanding indications, risks, benefits, and interpretation of cardiac catheterisation.
• Understanding physiology and pathophysiology of pulmonary hypertension including, hemodynamics and ventricular function, including familiarity and experience with Eisenmenger patients.
• Knowledge of classification systems for pulmonary hypertension.
• Comprehensive knowledge of all licensed drug classes for pulmonary hypertension, mechanism of action, evidence base, side effect profile, and drug interactions.
• Experience of initiating and monitoring patients on each class of therapy. Preferably experience in a variety of modes of delivery.
• Indications and contraindications for pulmonary vasodilator drugs.
• Knowledge of methods for assessing disease severity including clinical, hemodynamics, exercise testing, imaging, biomarkers.
• Recognition of deterioration, lack of improvement, and indications for treatment escalation.

Table 2. Recommendations for the final assessment of advanced training level in paediatric pulmonary hypertension

<table>
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<tr>
<th>Requirement</th>
<th>Example</th>
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<td>Log and/or direct observation of at least 50 assessments for suspected pulmonary hypertension (including inpatient and outpatient visits, at least 10 in intensive care). Besides the default diagnostic tools, these assessments should include also ventilation-perfusion scans and cardiac MRI where applicable.</td>
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<td>Log and/or direct observation of initiation of pulmonary arterial hypertension therapy in treatment naive patients: at least 5x parenteral prostacyclin analogues, 20x either endothelin receptor antagonists or phosphodiesterase inhibitors, or combination thereof.</td>
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<td>Log and/or in-training comprehensive interpretation of at least 50 invasive hemodynamic studies in the cardiac catheterisation laboratory (including at least 10 cases performed by trainee).</td>
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<td>Log and/or direct observation of at least 100 transthoracic echocardiograms in patients with pulmonary hypertension, including at least 20 neonates, 20 infants, and 20 children with CHD.</td>
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<td>Log and conductance of 6 minute walk testing in at least 20 children/young adults with pulmonary hypertension.</td>
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<tr>
<td>Log and/or direct observation of at least 20 cases of cardiopulmonary exercise testing in patients with pulmonary hypertension.</td>
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The numbers outlined in Table 2 are suggested as minimal requirements for final assessment of advanced level training.

The care of children with pulmonary hypertension is a rapidly evolving field that requires in-depth familiarity with new developments in physiology, pharmacology, and different diagnostic modalities, among others. Thus, an active contribution of the trainee to clinical and/or basic research is desirable, ideally documented by publication(s) in a peer-reviewed journal and/or presentation(s) at scientific meetings relevant to the field. In any case, the subspecialty fellow should aim to attend at least one specialty international meeting a year. A dedicated research period of 3-6 months (within a 24 months fellowship) is recommended for fellows working at academic centres, with the goal to produce a hypothesis-driven original research article as one of the lead authors. Research projects which were completed before the commencement of advanced training in pulmonary hypertension can be considered equivalent. Establishing a scientific advisory committee for each fellow is recommended.
• Recognition and treatment of complications in patients with pulmonary hypertension; pulmonary hypertension crisis, syncope, arrhythmia, thrombosis, right heart failure, pregnancy, thyroid disease.
• Supportive care of patients with pulmonary hypertension during intercurrent illnesses/undergoing invasive procedures for example, pneumonia, elective surgery, anaesthesia.
• Indications, risks, and benefits of atrial septostomy, and reverse Potts shunt.
• Indications, risks, benefits referral, and timing of lung and heart-lung transplantation.
• Indications, risks, benefits of short, and long-term mechanical support, including extracorporeal membrane oxygenation and ventricular assist device support.
• Outcomes of pulmonary hypertension nationally and internationally, including mortality and major morbidities.
• Understanding role of rehabilitation, psychology services, and palliative care.
• Familiarity with research areas and methodology relating to pulmonary hypertension management. This includes physiology, epidemiology, novel pharmacological treatments, and clinical trials of novel therapies.

Skills
• Counselling families with new diagnosis of pulmonary hypertension.
• Contributing to genetic counselling relevant to pulmonary hypertension led by an appropriately accredited genetics team.
• Eliciting a history and examination to aid diagnosis and assess treatment response.
• Assessment of ventricular afterload and function clinically and with imaging modalities.
• Interpreting and performing diagnostic cardiac catheterisation with acute vasodilator testing in patients with confirmed or suspected pulmonary hypertension, including comprehensive knowledge of cardiac catheterisation and acute vasoreactivity testing for decision making on (1) repair of CHD in the presence of elevated pulmonary vascular resistance, (2) idiopathic or heritable pulmonary arterial hypertension.
• Counselling about pregnancy risks and options for contraception in adolescents.

Attitude
• Ability for lifelong learning.
• Dedication to continued improvement in patient care through research, education, and evolving practice.

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