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CME article

Managing End Stage Lung Disease in Children

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EDUCATIONAL AIMS

- To illustrate the benefits of integrating of palliative care principles with restorative and life-prolonging care for children with End Stage Lung Disease (ESLD)
- To discuss the trajectory of ESLD in children and when to initiate Integrated Palliative Care Planning
- To review strategies applicable to the symptomatic and restorative management of ESLD
- To illustrate aspects of the psychological and social challenges faced by children and their families in coping with ESLD
- To discuss the impact of lung transplantation upon the management of ESLD

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SUMMARY

Over the course of a career most physicians will manage only a handful of children through End Stage Lung Disease. Nonetheless, the approach of the physician to this challenge will have a profound impact on the children and families they encounter. Managing the end of life well can bring personal growth and professional satisfaction. In this review we highlight aspects of the Palliative Care approach and its integration with restorative and life-prolonging care. We review the role of active treatment, respiratory support, symptom management and psychosocial aspects of the management of End Stage Lung Disease.
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INTRODUCTION

End Stage Lung Disease (ESLD) is uncommon in children. The prevalence of life limiting conditions in childhood has been estimated at 32 per 10,000 children in the UK from birth to 19 years of age.[1] Respiratory diagnoses account for 8.8–12% of the burden of life limiting conditions of childhood.[1,2] When ESLD is encountered it is usually in the context of Cystic Fibrosis or Broncho-Pulmonary Dysplasia. The scope of life-limiting lung conditions in childhood includes: bronchiectasis, interstitial lung diseases, surfactant deficiencies and conditions of the vasculature such as idiopathic pulmonary hypertension. It should be noted that mortality usually proceeds through a final common pathway of cardiorespiratory insufficiency and as such respiratory failure is a common feature of the end of life due to diverse disease processes in children.

The evolution of life-threatening lung disease in children can be quite variable, making a standard approach difficult. The Association for Children's Palliative Care (ACT) describe four patterns of evolution in life limiting conditions of childhood (Table 1).[3] It is clear from the variation in natural history of evolution that an individualised approach to management of ESLD will be necessary.

Emerging respiratory failure becomes evident in a child when their ability to accommodate stresses (respiratory infection, recovery from general anaesthesia, exercise, sleep) becomes limited. Aggressive optimisation of the medical management of lung disease is warranted including: extended microbiological investigation and tailored antimicrobial treatment, optimisation of airway clearance techniques, targeted efforts to improve nutritional status and, revisiting disease modifying strategies to retard underlying disease mechanisms.

As disease progresses, the waxing and waning nature of respiratory failure at low lung function can often make short term prognostication difficult even if the final outcome is clear. Signs that lung disease is reaching End Stage are described in Table 2.[4,5]

PALLIATIVE CARE APPROACH

At the end stages of lung disease, increasing support is often needed for symptom relief and to address the spiritual, social and

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Table 1
The Association for Children's Palliative Care describe 4 patterns of evolution of life limiting illnesses.

ACT Categories of Life Limiting Conditions (3)
1. Life-threatening conditions for which curative treatment may be feasible but can fail
2. Premature death is inevitable. There may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities.
3. Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years.
4. Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death.

psychological needs of the child and family. Active disease treatment often continues, but decisions are more often framed in the context of balancing treatment burden against benefit or treatment goals. The American Thoracic Society recommends that palliative care begin when a patient becomes symptomatic, should run concurrent with restorative and life-prolonging care, and be titrated to the needs of the patient and family.[6]

The Center to Advance Palliative Care have recommended "automatic" and "suggested" criteria for referral of paediatric pulmonology patients to a palliative care specialist. Their "automatic" criteria include: CF patients considering/at the time of lung transplant; CF patients with FEV_1<30%, ventilator dependence or ineligibility for lung transplant in CF patients, and, Bronchiolitis Obliterans. "Suggested" criteria include: CF patients with multiple hospitalizations, pain, dyspnoea or likely to benefit from symptom management, Central hypoventilation syndromes, and patients who are chronically ventilator dependent.[7]

The Palliative care approach has been defined as:

> '*An active and total approach to care, from the point of diagnosis or recognition, throughout the child's life, death and beyond. It embraces physical, emotional, social and spiritual elements and focuses on the enhancement of quality of life for the child/young person and support for the family. It includes the management of distressing symptoms, provision of short breaks, and care through death and bereavement.*'[3]

INTEGRATED PAEDIATRIC PALLIATIVE CARE PLANNING

Care planning and discussion is best initiated at the earliest stage when it is recognised that the child's lung disease is life threatening. This can be introduced to the family and child as a 'Preparatory Phase'. Barriers to addressing the issue may include the anticipation of transplant, unrealistic expectation of cure and family denial.[8] Care planning incorporates sensitive and regular provision of honest and realistic information about health status, clarity about the objectives of treatments, and structured discussion and documentation of the patient and family's wishes regarding options for care.[4] Plans are revisited and adapted as the child's and family's need evolves. Himelstein et al. recognise five practice spheres to be addressed in care planning (Figure 1).[9] More detailed advice on Paediatric Palliative care service planning can be found in the RCPCH(UK) Guide to the Development of Children's Palliative Care Services.[10]

Table 2
End Stage Lung Disease characteristics.

End Stage Lung Disease Characteristics (4, 5)
Persistent dyspnoea despite optimisation of medical management
Inability to maintain metabolic compensation for chronic respiratory acidosis
Decreased mobility
Increasing hospitalisation for chest infection or respiratory decompensation
Resistant respiratory pathogens
Limited improvement following hospital admission/prolonged intravenous antibiotic therapy
Accelerated rate of decline in pulmonary function despite therapy
Oxygen dependence
Pulmonary hypertension
Unrelenting weight loss that cannot be halted or reversed by supplemental nutrition

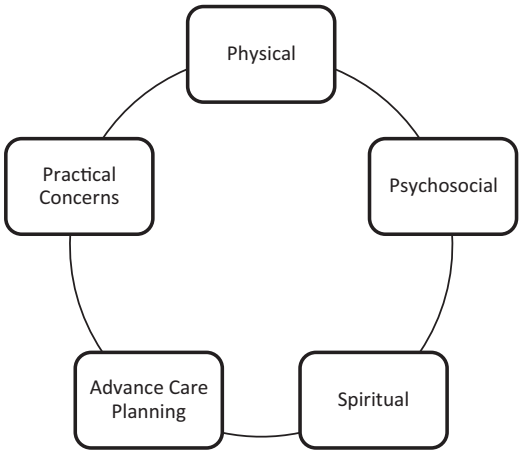

Figure 1. Core elements of Integrated Paediatric Palliative Care Planning (9).

LUNG TRANSPLANTATION

One of the most significant treatment decisions in ESLD surrounds lung transplant as a potential active treatment option. Important issues to consider include: treatment goals, suitability for transplantation, organ availability, timing of referral, potential survival gains, and use of aggressive treatment modalities designed to prolong life whilst waiting for a lung transplant.

The goal of lung transplant is to prolong life. For most patients it is a palliative rather than curative treatment. The process of assessment for lung transplantation can be stressful and involve prolonged separation from support networks. The burden of treatment is high following lung transplantation, and this must be factored into any consideration of the balance between survival and quality of life.[11]

Not all candidates will be eligible for transplantation. The ISHLT statement[11] defines absolute and relative contraindications and transplant centres can be consulted for advice. It is recommended that listing for transplantation occur when life expectancy is greatly reduced but nonetheless greater than the expected waiting time for a suitable organ. Transplantation should be performed when life expectancy after transplantation exceeds life expectancy without the procedure.[11]

Organ availability and allocation practices vary internationally. Algorithms may be based on waiting time, or the balance between waiting list survival and post-transplant survival for each candidate. In the US in 2006, 54 paediatric candidates received lung transplants and 16 died while waiting.[12]

The lung transplant half-life (i.e. the time after which 50% of paediatric lung transplant recipients have died) reported by the ISHLT for the period January 1990 to June 2009 was 5.5 years. The functional status of surviving paediatric lung transplant recipients is very good, with 86% of children having no physician-reported activity limitation even 5 years after transplant. The most common causes of late death after transplant are bronchiolitis obliterans and infection.[13]

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