

Pediatric Thoracic Organ Transplantation

Current Indications, Techniques, and Imaging Findings



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KEYWORDS

- Pediatric lung transplantation • Heart transplantation • Acute rejection • Chronic rejection
- Infection • Posttransplant lymphoproliferative disorder (PTLD)

KEY POINTS

- Accurate and timely radiologic diagnosis of transplant complications facilitates appropriate treatment and minimizes morbidity and mortality.
- As in adults, the major complication affecting long-term survival in pediatric lung transplant recipients is bronchiolitis obliterans from chronic lung rejection.
- The clinical and imaging features of post-lung transplant complications can overlap. Thus, knowledge of the time point at which the complication occurs is essential to distinguish the different entities.
- Allograft rejection in post-heart transplant patients remains one of the main complications limiting long-term graft survival and is the primary cause of death during the first 3 years posttransplant.
- Cardiac magnetic resonance (MR) imaging can be useful as a less invasive alternative to endomyocardial biopsy in the surveillance of acute cellular rejection after orthotopic heart transplant in conjunction with clinical and laboratory findings.

INTRODUCTION

Since the initial use of lung and heart transplantation for end-stage cardiopulmonary disease in the 1980s, substantial advancements have been made in surgical technique, immunosuppressive regimens, recognition and treatment of allograft rejection, and multidisciplinary long-term care.¹ Imaging is an essential component in the

evaluation of pretransplant and posttransplant pediatric patients for initial diagnosis, follow-up, assessment, and detection of complications. Clear knowledge of the spectrum of disease patterns and their relation to the time course from transplantation is essential for the appropriate management of these pediatric patients, particularly those presenting with acute rejection. Furthermore, recognition of characteristic imaging

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features is important because it can guide treatment and may obviate unnecessary additional imaging studies or invasive procedures such as biopsy or surgery. This article reviews the current indications and up-to-date imaging techniques for evaluating children both before and after thoracic organ transplantation and the spectrum of imaging findings that can occur in this patient population.

LUNG TRANSPLANTATION

Current Indications

Lung transplantation in the pediatric population should be considered in carefully selected patients who present with untreatable end-stage or progressively worsening advanced lung disease or pulmonary vascular disease for which there is no further available medical therapy. The common indications for lung transplant in children vary by age group (Table 1). Most pediatric patients who receive lung transplants are those with severe advanced cystic fibrosis. Another common diagnosis leading to lung transplantation in children is pulmonary hypertension, either idiopathic or related to congenital heart disease (CHD). In addition, less frequent but also important indications for transplant unique to children include pediatric interstitial lung diseases (eg, congenital surfactant deficiency syndrome and chronic lung disease of

infancy), congenital cardiac diseases involving the pulmonary vasculature, and primary pulmonary vascular conditions.

Bilateral lung transplantation is the most common surgical technique in children, especially in patients with cystic fibrosis, and is preferable in children with pulmonary hypertension.² Absolute contraindications for lung transplantation are mostly derived from experiences with the adult population and include active malignancy, sepsis, active infection such as tuberculosis, severe neuromuscular disease, refractory nonadherence, multiorgan dysfunction, and hepatitis C infection with histologic liver disease.³ Relative contraindications for lung transplantation include congenital or acquired immunodeficiency syndromes, renal insufficiency, poorly controlled diabetes mellitus, active collagen vascular disease, and severe scoliosis.³

Although short-term (1 year) survival rates for pediatric lung transplantation recipients have improved, long-term outcomes for children receiving lung transplantation have not. At present, survival in pediatric patients at 1 year after lung transplantation is approximately 80% and at 5 years approximately 50%.⁴ The most common cause of early mortality is related to primary graft failure, whereas the most common cause of death within the first year is infection.⁵ As in adults, the major complication affecting long-term survival is bronchiolitis obliterans (BO) from chronic lung rejection.⁵

Table 1
Common indications for pediatric lung transplantation by age

Age Group (y)	Indication for Transplant
<1 (infants)	<ul style="list-style-type: none"> • Surfactant protein B deficiency • Congenital heart disease • IPAH
1–5	<ul style="list-style-type: none"> • IPAH • IPF • Pulmonary fibrosis (other) • Retransplant
6–10	<ul style="list-style-type: none"> • CF • IPAH • Bronchiolitis obliterans, nonretransplant • Retransplant • IPF
11–17	<ul style="list-style-type: none"> • CF • IPAH • Retransplant

Abbreviations: CF, cystic fibrosis; IPAH, idiopathic pulmonary arterial hypertension; IPF, idiopathic pulmonary fibrosis.

Imaging Techniques

Imaging evaluation is essential in assessing the degree of existing lung disease and potentially ascertaining the underlying cause of the patient's respiratory dysfunction. It also plays an important role in managing the other stages of pediatric lung transplant patients, including assessing donor quality and size, and posttransplant monitoring.

The preoperative assessment of potential lung transplant recipients includes posteroanterior and lateral chest radiographs and computed tomography (CT) angiogram with two-dimensional (2D) and three-dimensional (3D) reconstructions of the airway and vascular structures. The combination of these imaging modalities is helpful in the assessment of the extent of the lung disorder, chest and lung size, mediastinal vessels, and large airways. Chest CT with multiplanar and 3D reformats is useful in detailing the anatomy of the airway and vascular structures before surgery. Ventilation-perfusion (V/Q) scans can provide information on lung function. V/Q scans also aid in

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