## American Society for Apheresis

## View Abstract

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TITLE: A BREATHTAKING SOLUTION: SIX YEARS OF ECP TO TREAT BRONCHIOLITIS OBLITERANS IN A PEDIATRIC LUNG TRANSPLANT RECIPIENT

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PRESENTATION TYPE: Oral or Poster

**CURRENT CATEGORY:** Extracorporeal Photopheresis

**ABSTRACT BODY:** 

**Purpose:** Extracorporeal Photopheresis (ECP) for chronic lung allograft rejection in the setting of bronchiolitis obliterans syndrome (BOS) is well-supported in the literature. However, the optimum duration is unknown. We report a case of a pediatric lung transplant patient with BOS whose lung function was improved and has been successfully maintained with chronic ECP for 5.6 years.

**Methods**: A male infant underwent a bilateral lung transplant at three months of age for Adenosine triphosphate (ATP) binding cassette subfamily A member 3 (ABCA3) mutation who developed chronic transplant rejection with BOS. He also developed chronic kidney disease (stage 3), iatrogenic adrenal insufficiency, osteopenia of multiple sites, and obesity. At the age of ten, due to a slow decline in FEV1, worsening hypoxemia, and pulmonary function below 30%, he was diagnosed with chronic allograft rejection with poor responsiveness to aggressive medical treatment. In keeping with ASFA Guidelines, he was considered a candidate for ECP (ASFA Category II Grade 1C evidence per the 2016 recommendations). ECP therapy was initiated with 2 sequential treatments every week for four weeks (induction therapy), spaced to two

treatments every two weeks for six weeks (consolidation therapy), and two treatments of ECP every month (maintenance therapy) thereafter. His lung function has improved and stabilized since initiation, with continued maintenance spanning 68 months (5.6 years).

**Results**: A chronic course of two treatments monthly has successfully maintained this patient's lung function over 68 months (5.6 years). The combination of ECP and standard immunosuppressive therapy stabilized and improved his FEV1 and his quality of daily life. He will continue monthly treatments and hopes to transition this regime into adult care.

Conclusion: In recent years, short-term outcomes after lung transplantation have improved due to advances in immunosuppressive regimens and better peri- and postoperative care. Long-term survival remains limited, however, with a median 5-year survival of approximately 54% and a median 10-year survival of 32%. Median survival after BOS diagnosis is 3 to 5 years. Since long-term survival after lung transplant is hampered by the development of chronic rejection, therapeutic strategies have been largely unsuccessful. Current practices primarily include conventional immunosuppression and traditional infectious prophylaxis. Our patient has been stable for 68 months (5.6 years) on ECP with improved pulmonary function since induction and good quality of life without incidences of recurrent acute rejection. Early initiation of ECP treatment may be a promising option for patients with solid organ transplants experiencing chronic rejection sequela. It could play an expanded role in helping to slow or prevent the decline of chronic rejection of solid organ transplant recipients and, more specifically, lung transplant patients experiencing BOS. We believe this is the longest reported case of lung transplant with BOS supported by chronic ECP in a pediatric patient.

## **DEGREE:**

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ASFA Membership (Abstract Submission): Yes, I am an ASFA member

ASFA Oral Presentation Awards (ASFA Members and Non-Members): Apply

ASFA Junior Investigator Abstract Award or ASFA Allied Health Abstract Award (ASFA Members Only):

ASFA Junior Investigator Abstract Award

(No Table Selected)



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