



TITLE: Lung transplantation for chronic pulmonary graft versus host disease in pediatric allogeneic hematopoietic stem cell transplant recipient

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ABSTRACT (up to 300 words)

A 13-year-old boy was diagnosed with Philadelphia-positive acute lymphoblastic leukemia at the age of 9. Considering the high-risk profile of his disease, a matched sibling donor hematopoietic stem cell transplantation (HSCT) was performed.

In the first 90 days, he did not show signs of acute GvHD. Around day + 240 he developed esophageal cGvHD. On day +350 lung cGvHD was confirmed. He was treated with steroids, cyclosporin, and mycophenolate mofetil. On day +442 chronic lung GvHD worsened, the patient developed subcutaneous emphysema, pneumothorax, and pneumomediastinum. A CT scan showed bronchiolitis obliterans and the lung function test showed restrictive and obstructive lung disease (FEV₁ 14 %). The imatinib treatment was introduced. Concomitant disease at this point was severe esophageal stenosis (diameter < 2 mm) demanding gastrostomy. Ruxolitinib treatment started. On day + 501. mini extracorporeal photopheresis started. Around day + 561. etanercept was introduced. In December 2021, he became oxygen dependent. In February 2022 he was presented to the National cGVHD team with the suggestion to proceed to a lung transplant. At that time, he was severely malnourished (BMI 11.5 kg/m², Z score – 4,82), with esophageal stenosis, which was a contraindication for a lung transplant.

In April 2022, he became SARS-CoV-2 positive.

This further deteriorated his lung function demanding high-flow nasal cannula respiratory support. Over time, he gained weight through continuous enteral feeding. Two international centers were consulted to proceed with lung transplantation, but he was refused. In August 2022, he developed ARDS demanding mechanical ventilation. In October 2022, the Croatian lung transplant team put the patient on the waiting list. Finally, in late December 2022, the patient had a lung transplant.

Eight months after the lung transplant our patient is in stable clinical condition with normal lung function.



BIOGRAPHY (up to 200 words)

Zrinko Šalek has completed his MD in July 2014 from Zagreb Medical School, Croatia. He finished his pediatric residency in November 2021 and works at the Division of pediatric hematology and oncology. His field of interest is pediatric hematology and oncology, more specifically hematopoietic stem cell transplantation in children and coagulation disorders. He is final year PhD student at the PhD study Biomedicine and Health Sciences in Zagreb. He has published several papers and co-authored in two university books. at School of Medicine University of Zagreb has published several scientific papers and authored two books. He is a tutor of the University subject fundamentals of medical skills at the School of medicine, Zagreb.

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