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**Successful sequential haploidentical maternal haematopoietic stem cell and kidney transplantation without requirement for long term immunosuppression**

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**Objectives :** Schimke Immuno-osseous Dysplasia (SIOD) is a rare autosomal recessive disease, occurring in 1 in 3 million caused by mutation of SMARCAL1 gene. Life expectancy mainly relies on management of end-stage kidney disease and prevention of recurrent and potentially fatal opportunistic infections.

**Methods :** A 5-year-old girl of non-consanguineous parents presented with SRNS secondary to focal and segmental glomerulosclerosis (FSGS), short stature, dysmorphism and normal development. Genetic analysis confirmed SIOD, and she commenced haemodialysis (HD) one year later. She had previously shown adequate vaccine responses, but further presented progressive T cell lymphopenia.

**Results :** Successful haploidentical haematopoietic stem cell transplantation (HSCT) followed by living related kidney transplantation (LRKT) from the same donor parent to allow tolerance of the allografts. She was switched from HD to CVVH at HSCT with pre-HSCT conditioning regimen, urothelium protection and infectious prophylaxis. She developed mucositis, fungal chest infection and cutaneous GVHD. She had stable blood pressure under CVVH with conversion back to HD on day +24 with 100% donor engraftment without systemic immunosuppression. She underwent successful high risk LRKT in view of vascular (small vessels requiring mesh), immunological and pre-transplant hypertensive risks from her mother 6.25 months after HSCT with expected zero HLA mismatch and negative crossmatch without induction or anti-proliferative agents. Her renal function normalised on day 5 with discontinuation of corticosteroids and tacrolimus by five weeks post-transplant. She has normal estimated glomerular filtration rate, absence of proteinuria and normal serum albumin with good T-cell reconstruction.

**Conclusions :** Apart from being the first child in the United Kingdom to undergo these sequential procedures, it is the first described case of SIOD patient undergoing HSCT under CVVH prior to future kidney transplantation. This successful intervention led to the LRKT with minimal immunosuppression and enabled better blood pressure control and increased quality of life for this patient.