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Double Umbilical Cord Blood Transplantation in a Pediatric Patient: A First for Nebraska

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Table 1.

Patient demographics and clinical characteristics.

	By Case (n=47) N (%)	By Patient (n=14) N (5)
Gender		
Male	17 (36.1)	5 (35.7)
Female	30 (63.8)	9 (64.6)
Age¹		
0-5	19 (40.4)	
6-10	6 (12.7)	
11-15	11 (23.4)	
16-20	8 (17)	
21+	3 (6.4)	
Average	Mean 10 y	
Clinical Characteristics		
Seizure history ²		5 (35.7)
Contractures		14 (100)
Photophobia		6 (42.8)
Able to ambulate		10 (71.4)
IQ³		
≤50		7 (50)
>50		6 (42.8)
Mean=55 Median=50.5		

¹Age at time of anesthetic, ²Includes current seizure disorder treated with medication, ³Stanford-Binet full scale IQ, 1 patient without documented IQ

with adherence of monitors and skin tearing upon removal of adhesives. Other complications were patient specific and not related to SLS.

Conclusion: Patients with SLS typically require general anesthetics when undergoing diagnostic studies and procedures for symptom management. Our case series suggests that general anesthesia is well tolerated in this population. Important aspects of pre-anesthetic evaluation include history of seizure severity and control, neurocognitive assessment and skin examination. Intraoperative considerations include photophobia, difficulty securing PIVs and monitors, relative heat intolerance, hypohydrosis, and challenges with positioning, padding, and line placement due to contractures. The ichthyosis of SLS spares the midface eliminating difficulty in securing airway devices. ■

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Double Umbilical Cord Blood Transplantation in a Pediatric Patient: A First for Nebraska

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Mentor: Sachit Patel

Program: Pediatrics

Type: Case Report

Background: Unrelated umbilical cord blood (UCB) transplantation has been used as a hematopoietic stem cell source for 30 years. Compared to adult bone marrow and peripheral blood stem cells, UCB has more rapid availability, absence of donor attrition, and reduced risk of GVHD despite HLA disparity. Unfortunately for larger patients, a single UCB unit has an insufficient amount of total nucleated cells to support engraftment. The use of two well-matched UCB units has been shown to overcome this barrier.

Methods: Chemotherapy, Graft, Cord Blood Transplant. Consent was obtained to utilize this case for educational purposes.

Results: A 12 year-old presented with bleeding and weight loss and was found to have anemia, thrombocytopenia and leukocytosis. Peripheral smear demonstrated auer rods. He was diagnosed with acute myeloid leukemia (AML-M4). He successfully completed therapy with protocol AAML 1031 but relapsed seven months later. He achieved a second complete remission with protocol AAML 0523. Given his high risk disease and poor prognosis with chemotherapy alone, the patient was offered transplant. No suitable sibling or adult unrelated donors were found. He underwent

mismatched double umbilical cord transplant. Complications included grade III aGVHD involving the skin and GI system, and CMV reactivation. Engraftment was achieved on day +27. He is now 16 years-old with full donor chimerism and complete immune reconstruction, no findings of chronic GVHD and no disease relapse.

Conclusion: This was the first pediatric patient in Nebraska to have a successful double UCB transplant. Double cord transplant is an acceptable alternative when there is no sibling or unrelated donor match and when a single cord unit total nucleated dose is insufficient. ■

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Shwachman-Diamond Syndrome: First Successful Hematopoietic Stem Cell Transplant in Nebraska

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Type: Case Report

Background: Shwachman-Diamond syndrome (SDS) is an autosomal recessive condition characterized by bone marrow dysfunction, pancreatic insufficiency, and

skeletal abnormalities. Ninety percent of patients with SDS have a mutation in the SBDS gene on chromosome 7, while the other ten percent are diagnosed clinically.