



Shriners Hospitals
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Pattern of Limb Involvement in Constriction Band Syndrome

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Objectives

- Constriction band syndrome (CBS) is a congenital limb anomaly with varying clinical presentation, and there is no clear diagnostic criteria.
- The etiology is unknown, and it is unclear which children are most at risk
- We sought to characterize patterns of limb involvement and potential demographic risk factors in CBS by analyzing a large cohort of patients.

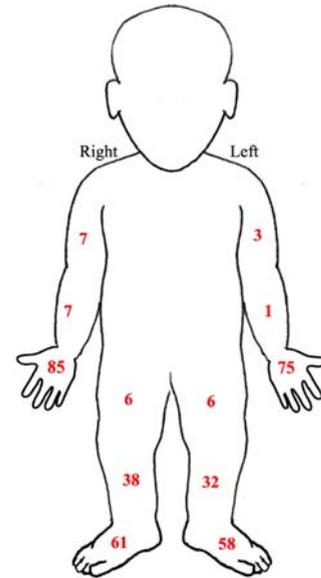
Methods

- We performed a retrospective review of all patients with CBS presenting to our tertiary medical center between 1998-2018.
- Patients were identified by ICD-9 and ICD-10 codes. Medical records were reviewed for demographic data and associated conditions. Demographics were compared to statewide population averages.
- Clinical photographs and radiographs were reviewed to determine the pattern of limb involvement.

Results

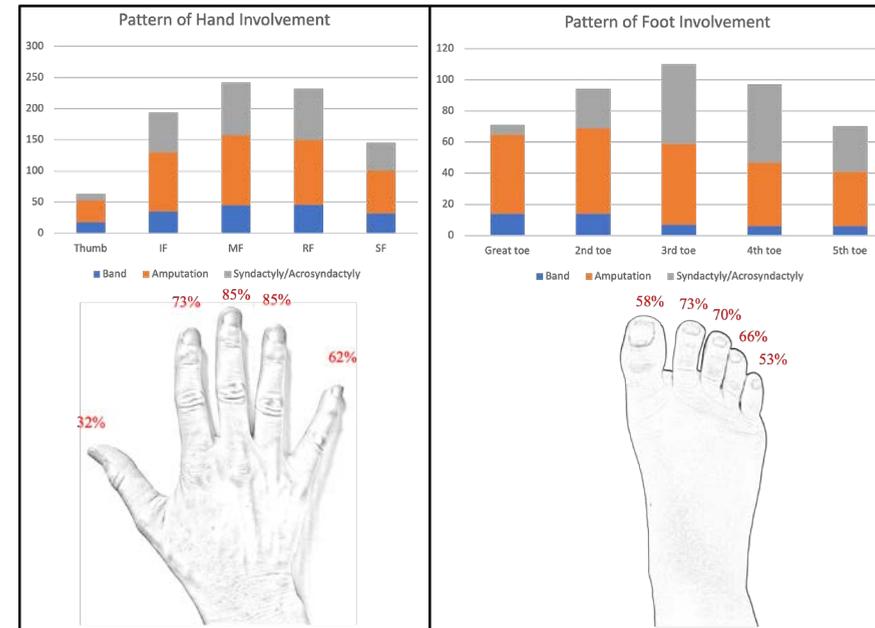
- 128 children with CBS were identified.
- The most prevalent feature was distal limb constriction bands (96%) followed by limb or digit amputations (88%) and syndactyly/acrosyndactyly (syndactyly with a proximal sinus) (69%). Amputations were characterized by presence of a terminal bone defect on radiographs but normal bone formation proximal to the amputation.
- The average number of involved extremities was 2.6 limbs per child, however 23% had involvement of only one limb.
- Children with at least one additional diagnosis had more limbs affected by CBS compared to children who were otherwise healthy (2, IQR 1-3 vs. 3, IQR 2-4, p=0.006), suggesting a more severe phenotype.

Location of limb involvement in 349 involved limbs



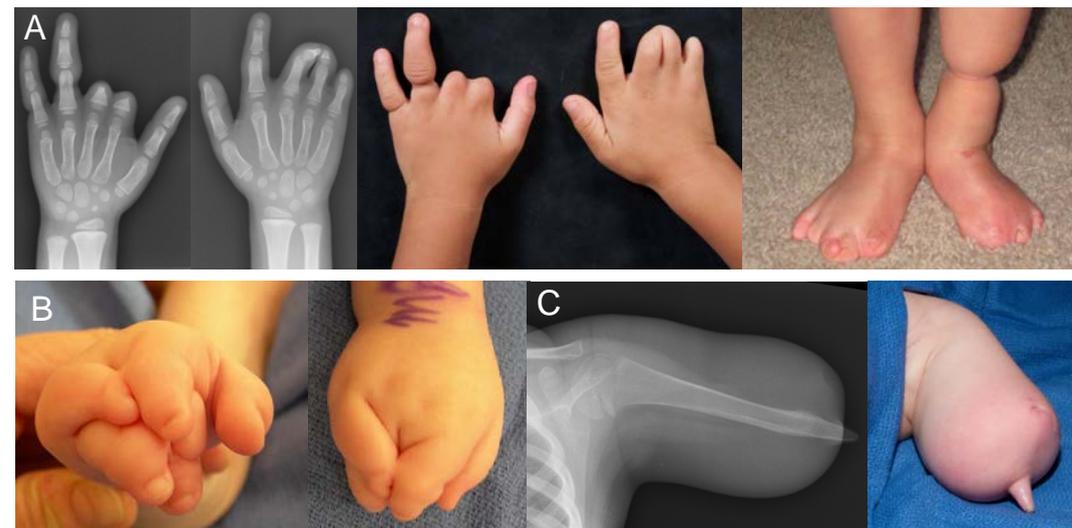
83% of children demonstrated upper extremity involvement and 80% had lower extremity involvement.

Location of digit involvement in 156 hands and 104 feet



There was a strong predilection for involvement of central digits of the hands and feet with sparing of the thumb/great toe and small finger/toe

Prevalent features



A. Child with amputations in all 4 extremities, constriction bands of the left small and ring fingers as well as the left ankle, and acrosyndactyly of the right middle and ring fingers.

B. Child with non-adjacent syndactyly and constriction band of the index finger.

C. A child with no constriction bands but demonstrate overgrowth of proximal limb amputations.

TABLE 1 Demographics and Prenatal Risk Factors

Patient characteristic	CBS (n=128)	CA newborn population average ^{15,16,17}	p-value
Male¹⁶	52%	52%	0.94
Maternal age at patient's birth¹⁷	25.6 years	28.3 years	<0.001
15-19	12%	6%	
20-34	68%	77%	
35+	20%	16%	
Paternal age at patient's birth	28 years	-	-
Race/Ethnicity¹⁶			
White	47%	45%	0.81
Black	10%	9%	0.89
Asian	10%	16%	0.03
Hispanic	25%	29%	0.21
American Indian/Alaska Native	9%	-	-
Language¹⁶			
English	87%	77%	0.002
Other	13%	23%	0.002
Insurance¹⁶			
Private	33%	37%	0.32
Public	57%	58%	0.83
Uninsured	10%	6%	0.12
Area Deprivation Index¹⁵			
National percentile	39%	50%	<0.001
State decile	7.5	5	<0.001
Gestational trauma	43/112 (38%)	-	-
Premature¹⁶	49/106 (46%)	9%	<0.001
Low Birth Weight¹⁶	19/67 (28%)	7%	<0.001
First born	36/84 (43%)	-	-

CBS was associated with young and old maternal age, gestational trauma, prematurity, low birth weight, and high social deprivation compared to state averages.

Results cont.

- Children with more limbs involved underwent more surgeries than those with fewer limbs involved (p<0.001).
- The most common associated conditions were clubfoot (34%) and craniofacial anomalies (12%).

Conclusions

- Children with CBS can be categorized as having either mild or extensive involvement based on the number of involved limbs and associated conditions.
- Future investigation of prenatal risk factors is necessary to further elucidate the etiology of this heterogenous condition.