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Title

Clinical Manifestations of Constriction Band Syndrome

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INTRODUCTION

- Constriction band syndrome (CBS) comprises a heterogeneous collection of congenital anomalies that affect the extremities. Infants present with constriction bands causing skin indentations, limb amputations, and syndactyly.
- Due to the highly variable presentation, there are currently no clear diagnostic criteria. A standardized diagnostic criteria would help physicians distinguish CBS from other terminal deficiencies

OBJECTIVES

- Characterize the clinical manifestations of CBS by retrospectively analyzing a large cohort of patients
- Use this data to propose diagnostic criteria to standardize the diagnosis of CBS and help differentiate CBS from other congenital limb conditions
- Evaluate possible risk factors for CBS

METHODS

- Retrospective chart review of all children with CBS presenting at our tertiary care hospital. Patients were identified via ICD 9/ICD 10 codes.

Inclusion criteria

- Treated between January 1, 1998 and December 31, 2018
- Clear, detailed description of clinical findings by a pediatric orthopedic surgeon
- Presence of one or more pathognomonic findings:
 - Constriction bands
 - Acrosyndactyly (syndactyly with a proximal sinus)
 - Non-adjacent syndactyly
 - Bony overgrowth of an amputated limb or digit

- Pattern of limb involvement was aggregated from clinical notes, photographs, radiographs. Associated diagnoses, demographics, and birth history were collected from the medical record

Data Analysis

- Basic statistics (mean, SD) completed for demographics
- Bivariate statistical analysis to assess whether non-CBS diagnoses were associated with the severity of limb involvement; to evaluate the demographics and prenatal histories of patients with CBS differ from those of the general population

- Student's t test used for parametric continuous variables and Mann-Whitney U test and Kruskal-Wallis for nonparametric continuous variables

RESULTS

TABLE 1 Characterization of limb involvement and clinical presentation (n=128)

Feature	Frequency (%)
Upper extremity involvement	83%
Lower extremity involvement	80%
Constriction band	96%
Limb or digit amputations	88%
Syndactyly/acrosyndactyly	69%
Associated diagnosis	52%
Clubfoot	34%
Craniofacial anomalies	12%
Genitourinary abnormalities	6%
Cleft palate	3%
Cardiac anomalies	3%

- The average number of involved extremities was 2.6 limbs per child. 23% of children had involvement of only one limb.
- Children with at least one additional diagnosis had more limbs affected by CBS than those who were otherwise healthy (2, IQR 1-3 vs. 3, IQR 2-4, p=0.006)



FIGURE 1 Pathognomonic findings of CBS

FIGURE 2 Central digits were affected more frequently than peripheral digits

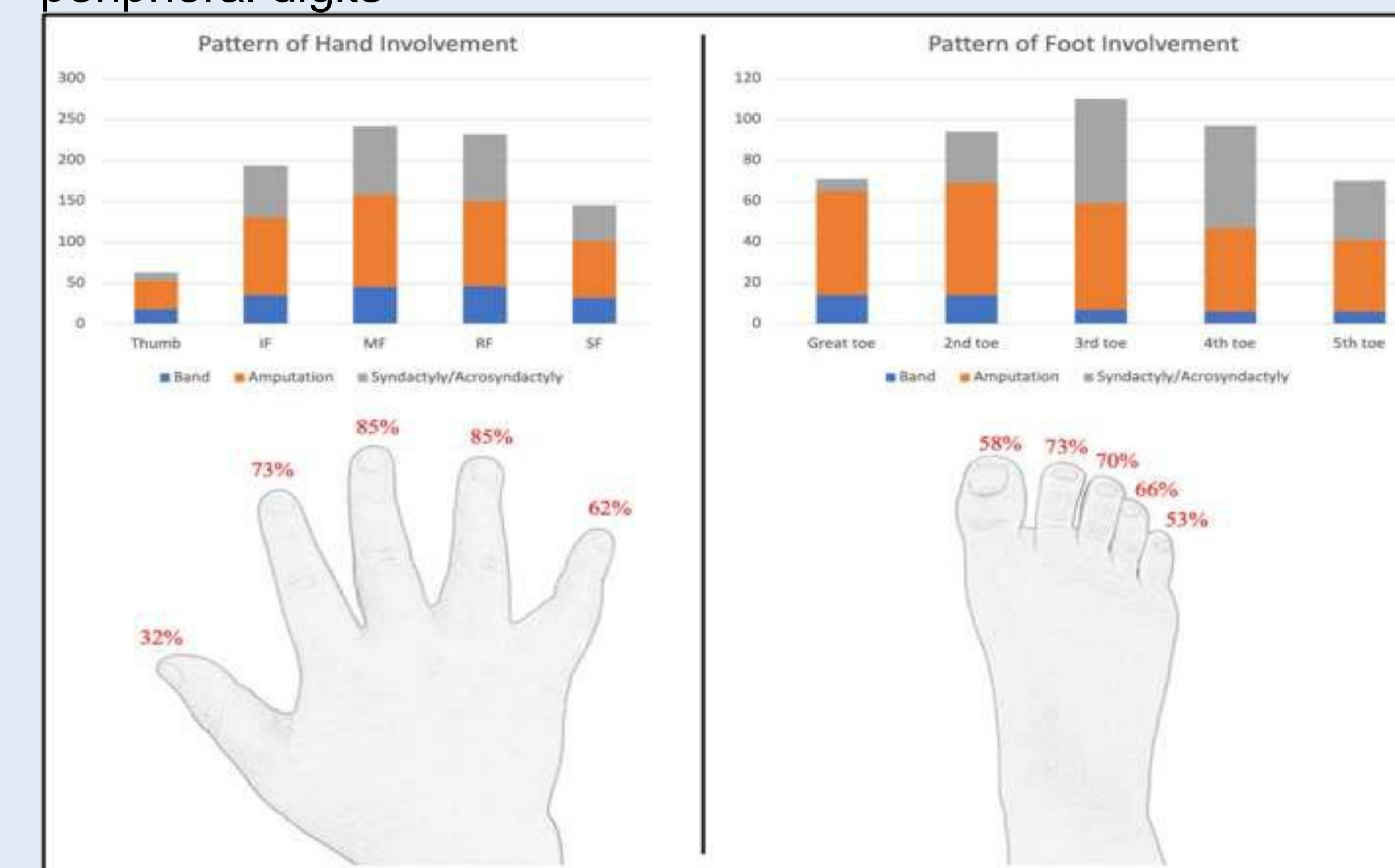


TABLE 2 Demographics and Prenatal Risk Factors

Patient characteristic	CBS (n=128)	CA newborn population ²⁻⁴	p-value
Male³	52%	52%	0.94
Maternal age at patient's birth⁴	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/Ethnicity³			
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%	-	-
Language³			
English	87%	77%	0.002
Other	13%	23%	0.002
Insurance³			
Private	33%	37%	0.32
Public	57%	58%	0.83
Uninsured	10%	6%	0.12
Area Deprivation Index²			
National percentile	39%	50%	<0.001
State decile	7.5	5	<0.001
Gestational trauma	43/112 (38%)	-	-
Premature³	49/106 (46%)	9%	<0.001
Low Birth Weight³	19/67 (28%)	7%	<0.001
First born	36/84 (43%)	-	-

DISCUSSION

Proposed diagnostic criteria

- Presence of one or more pathognomonic findings: constriction bands, acrosyndactyly or non-adjacent syndactyly, and bony overgrowth of an amputated limb or digit (Figure 3)
- If congenital amputations are present, the bones proximal to the level of amputation are usually normal (not malformed or dysplastic)

Risk Factors

- We found high rates of gestational trauma, prematurity, and low-birth weight, suggesting intrauterine trauma may play a role in CBS.
- Maternal age was frequently at the extreme ends of the spectrum (≤ 19 and ≥ 35 years old), which are known to be associated with pregnancy complications.⁴
- Children with CBS were significantly more disadvantaged than the state average. This supports previous research suggesting low economic status may be a risk factor for CBS.⁵⁻⁷
- Further research of prenatal risk factors is needed.

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