Congenital constriction ring syndrome: Observations, surgical approach and optimization of treatment

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Introduction

Constriction ring syndrome (CRS) is a congenital condition with an incidence of 1:2000 to 1:15,000 live births. Upper limbs and distal extremities are most frequently affected. It can present with complex deformities ranging from constriction grooves in skin to complete amputation, more commonly affecting the upper limb and distal extremities. Patterson classified these deformities into simple ring constrictions, constrictions with distal deformity (with or without lymphoedema), constrictions accompanied by fusion of distal parts and uterine amputation.

We report our experience in managing this complex condition.

Method

A retrospective review of children with CRS referred to a specialist centre was performed. Patient demographics, anatomy of constriction, Patterson classification, management and outcome were recorded.

Results

23 patients were studied. (M:F ratio 11:12). 22 (96%) had upper limb involvement of which 11 (48%) were bilateral. 5 (22%) were born prematurely, 5 (22%) had other congenital anomalies and 9 (39%) had lower limb involvement also.

Patterson Classification

n=33 (upper limb involvement only)

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Number</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>Simple ring constriction</td>
<td>5</td>
</tr>
<tr>
<td>II</td>
<td>Ring constriction + deformity +/- lymphoedema</td>
<td>2</td>
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<tr>
<td>III</td>
<td>Ring constriction + fusion distal parts</td>
<td>14</td>
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<tr>
<td>IV</td>
<td>Uterine amputation</td>
<td>12</td>
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Deformities observed:

- Constriction ring
- Digits – distal lymphoedema
- Upper limb – skin and deeper
- Acrosyndactyly
- Amputations (multiple levels seen)
- Complex combinations of the above

Discussion

Specific aspects of our treatment approach include optimization of skin condition at each stage of treatment and excision of constriction rings on limbs without z-plasty using longitudinal fasciotomies. Not all patients require surgery but consider urgent surgical intervention if there is evidence of nerve or vascular compromise.

The Patterson classification is a helpful guide, but in reality there is often a complex combination of deformities in multiple digits at multiple levels. Appropriately timed staged surgical intervention maximizes function in these complex patients. Functional and aesthetic improvement was observed in most patients.

Surgical Intervention

18 / 23 underwent surgery, with 12 requiring multiple/staged procedures. Procedures included digital separation, release of acrosyndactyly, web deepening, stabilization of distal lymphoedematous digit, excision of constriction rings and debulking of lymphoedema. The mean age at surgery in our cohort was 18.7 months of age. Optimization of skin condition at every stage is advocated.

2 cases required urgent surgery:

(i) constriction ring excision, longitudinal fasciotomies and neurolysis on the upper limb in a premature infant with a high median and ulnar nerve palsy.

(ii) excision of distal lymphoedema of the hand.

Post-op: Staged surgery - excision of distal lymphoedema
Intra-op: Excision of constriction ring and neurolysis
Pre-op: Excision of distal lymphoedema
Post-op: Excision of constriction ring and neurolysis – return of motor function and well healed scar without z-plasty

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