Radial Longitudinal Deficiency

Radial Longitudinal Deficiency (RLD) is the current term used to describe the congenital anomaly previously known as Radial Club Hand. This change reflects recognition of the involvement of the whole upper limb. In RLD, the radial side of the upper limb is malformed, leading to various characteristic clinical features depending on the severity of the condition. The ulnar side of the limb is less or not affected in this condition. Ulnar Longitudinal Deficiency (ULD), where the anomalies affect the ulnar side of the limb also exists, but is much less frequently seen than RLD.

Introduction

Anatomy

The skeletal elements on the radial side of the limb, particularly the radius, are the most prominently affected, but the soft tissue elements are also affected in the condition. Lamb ([Lamb1977]) provides a brief description of the abnormal anatomy in RLD.

Associated Anomalies

Same Limb

The most important associated anomaly that requires surgical attention is the hypoplastic thumb, which is frequently present.

Other anomalies

- VATER
- VACTERL
- TAR
- Holt-Oram
- Fanconi Anemia

Classification

Bayne and Klug's classification is the most widely used. It is a radiological classification based on the severity of radial aplasia.

The modified Bayne and Klug has been proposed to include less and more severe forms of the condition. From the original types I-IV have been added N, 0 and V.

<table>
<thead>
<tr>
<th>Type</th>
<th>Anomaly</th>
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<tbody>
<tr>
<td>N</td>
<td>Isolated thumb anomaly</td>
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<tr>
<td>0</td>
<td>Deficiency of the carpal bones</td>
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<tr>
<td>I</td>
<td>Short distal radius ( (\text{delayed distal epiphysis}) )</td>
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<tr>
<td>II</td>
<td>Hypoplastic distal radius in miniature ( (\text{both distal and proximal epiphysis involved}) )</td>
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<tr>
<td>III</td>
<td>Absent distal radius ( (\text{absent distal or proximal epiphysis with very short radius}) )</td>
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<tr>
<td>IV</td>
<td>Complete absent radius</td>
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<tr>
<td>V</td>
<td>Complete absent radius and manifestations in the proximal humerus</td>
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</tbody>
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Natural History

General Review

Non-Operative Management

- Complements operative treatment
- starts with stretching exercises - wrist and elbow
- can splint/ cast - ? optimal regimen unknown
Surgical Technique

Takagi and colleagues have written a useful [https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5447903/][1] of the surgical technique of centralization. They also introduce a technique of radial lengthening for Bayne I and II RLD.

Outcomes

Centralization is Standard of Care:

Long-term results (Lamb):

1. Forearm still short 50-60%
2. Ulnar carpal fusion common
3. Risk of recurrent deformity
4. Risk of wire breakage

Further Reading

[Notes or in a formatted version](#) with working links