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Poly-Hill Sign and Popeye Arm in Facioscapulohumeral Muscular Dystrophy

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A 22-year-old man presented with gradually progressive proximal weakness and thinning of all four limbs for 5 years. There was no relevant family history. Neurological examination revealed facial weakness with an inability to blow, and a characteristic facioscapulohumeral pattern of muscle involvement with prominent shoulder-girdle and upper-arm wasting. A positive Beevor sign, reflecting preferential weakness of the lower abdominal muscles, was also present.

Posterior examination with shoulder abduction and elbow flexion demonstrated prominent scapular winging and a characteristic undulating contour of the shoulder girdle – the poly-hill sign (Fig. 1A). The scapulohumeral wasting was asymmetric and more pronounced on the left. On anterior inspection, there was marked wasting of the upper-arm musculature with relative preservation of the deltoids and forearm muscles, producing the characteristic "Popeye arm" appearance (Fig. 1B). Lower-limb examination demonstrated relative preservation of the calf musculature (Fig. 1C).

The combination of facial weakness, scapular stabilizer weakness, asymmetric humeral muscle wasting with relative deltoid and forearm sparing, a positive Beevor sign, and relative preservation of the calf musculature was characteristic of facioscapulohumeral muscular dystrophy (FSHD)[1-3]. Genetic confirmation could not be performed because of financial constraints.

FSHD is characterised by a selective and often asymmetric pattern of muscle involvement, typically affecting the facial and scapular muscles followed by the humeral and abdominal musculature[1,2,3]. Preferential wasting of the biceps and triceps with relative preservation of the deltoid and forearm musculature produces the characteristic

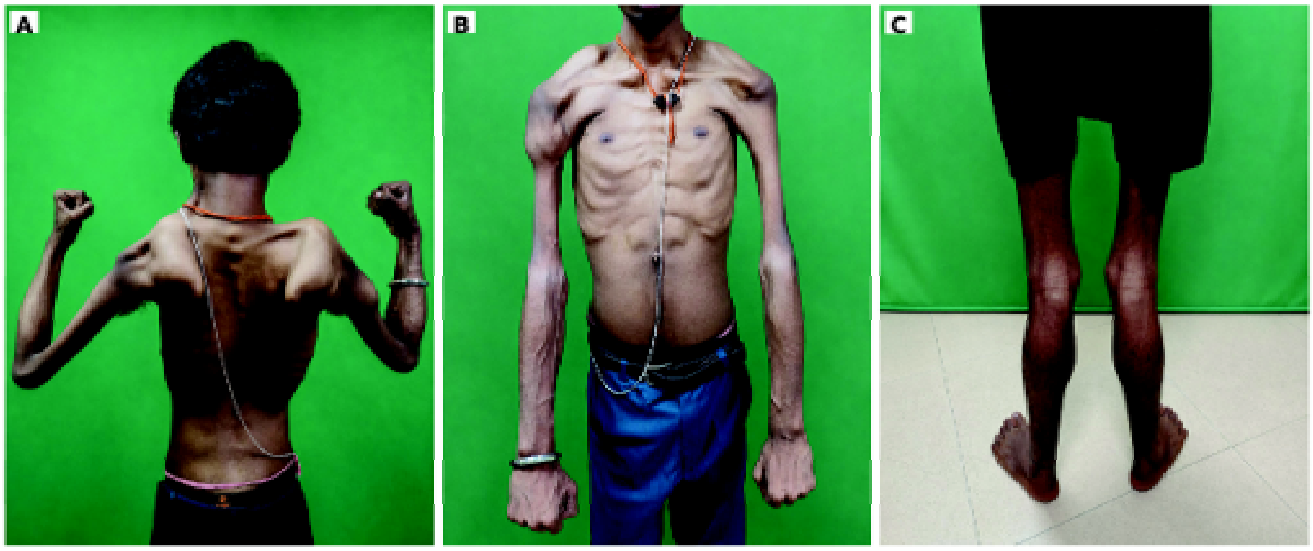


Fig. 1: *Characteristic clinical findings in facioscapulohumeral muscular dystrophy.*

A: Posterior view with shoulder abduction and elbow flexion demonstrating asymmetric scapular winging and the characteristic poly-hill sign, produced by selective scapulohumeral muscle wasting with relative preservation of portions of the deltoid. The muscle wasting is more pronounced on the left side.

B: Anterior view demonstrating asymmetric humeral muscle wasting, more marked on the left, with relative preservation of the forearm musculature, producing the characteristic "Popeye arm" appearance.

C: Posterior view of the lower limbs demonstrating relative preservation of the calf musculature.

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Popeye arm. Differential involvement of the scapular stabilizers with relative preservation of portions of the deltoid and prominence of underlying bony landmarks produces the alternating elevations and depressions of the poly-hill sign. A positive Beevor sign, resulting from preferential lower abdominal muscle weakness, is another useful bedside clue[2,3].

Recognition of these characteristic bedside signs can provide an important diagnostic clue to FSHD, particularly where molecular testing is unavailable or inaccessible.

References

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