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Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

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Dear Editor,

Joubert's syndrome is indeed a rarity in clinical and radiological practice with a myriad of accompanying anomalies.

Herein we present a case of an eight-month-old female infant who presented with delayed milestones, nystagmus and post-axial polydactyly (Figure 1). She was referred for MRI of the brain after a suspicious neurosonographic

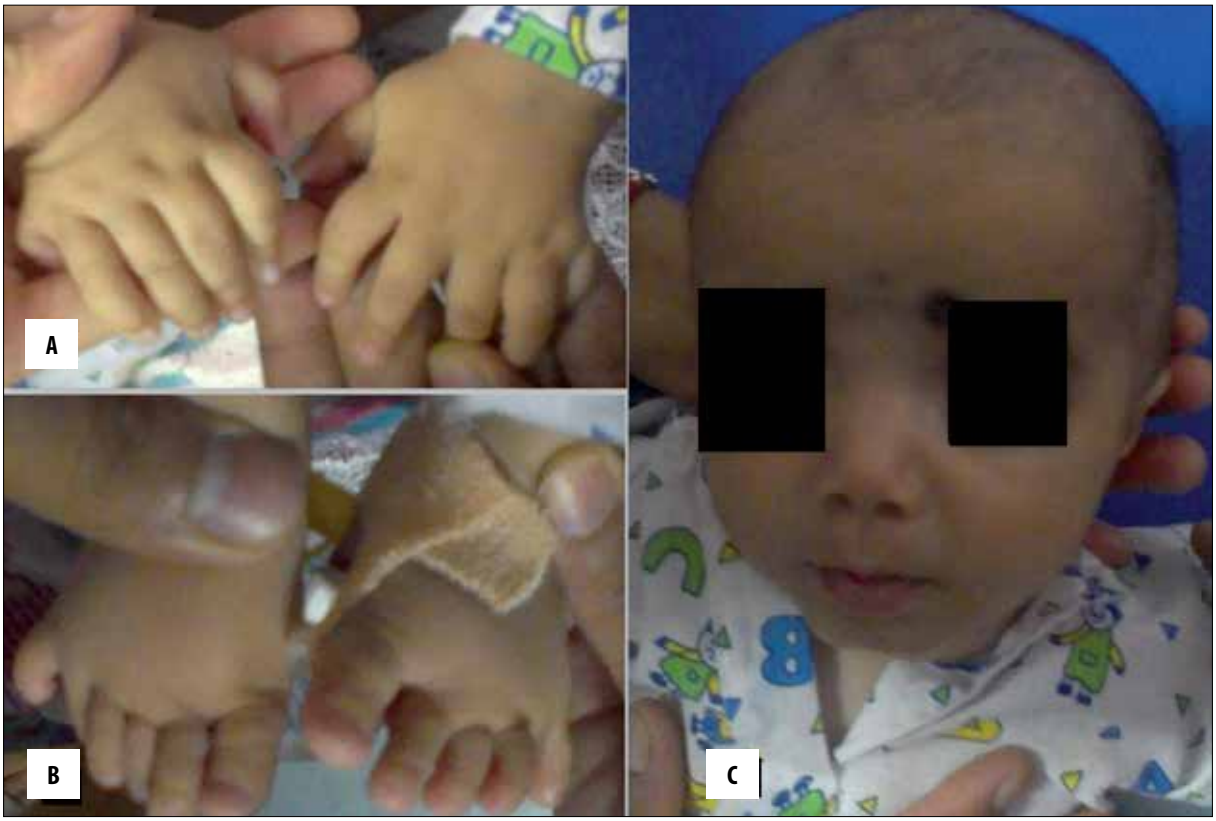

Figure 1. Clinical photos of the patient showing post-axial polydactyly in the upper and lower limbs (**A, B**) and broad flat nasal bridge (**C**).

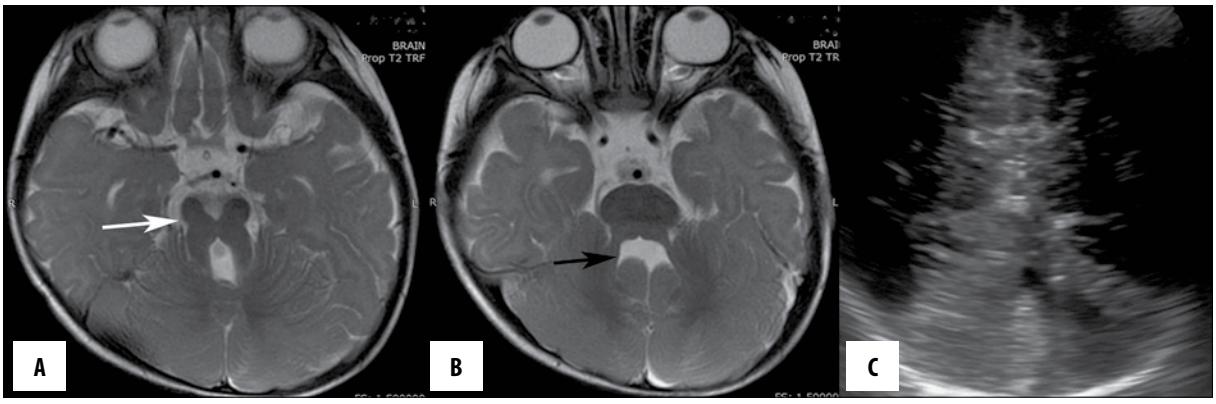

Figure 2. Axial T2-weighted MR images (**A, B**) showing typical molar tooth appearance of the midbrain (white arrow in **A**) and batwing appearance of the fourth ventricle (black arrow in **B**). Oblique coronal neurosonogram (**C**) showing a poorly visualized cerebellar vermis.

finding of poorly visualized vermis. Brain MRI revealed classical findings of Joubert syndrome, i.e. molar tooth appearance of the midbrain and batwing-shaped fourth ventricle (Figure 2).

In addition to the described clinical picture, oculofacial and digital anomalies, as well as neuroradiological association of cortical dysplasias, grey matter heterotopias, ventriculomegaly, and corpus callosum agenesis [1], certain other radiological associations are to be looked for.

Other described findings include hippocampal malrotation (unilateral and bilateral), occipital cephalocele, absent septum pellucidum, dysmorphic brainstem (especially mesencephalon and tectum), atrophic pons, interpeduncular heterotopias, delayed myelination, deep foramen cecum, thickened or small cerebellar peduncles, absence of flocculonodular lobe (rarely), rostral shift of the central velum fastigium and polymicrogyria [2–4]. Association of Joubert syndrome with tectocerebellar dysplasias or Dandy-Walker malformation has been labeled as Joubert syndrome plus [3]. Very recently, an additional neuroimaging sign, the so-called 'shepherd's crook sign', has been described [5].

In conclusion, when encountering a case of Joubert syndrome, assessment of the above described associated anomalies is worthwhile on the same scan.

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