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**Zimmerman-Laband Syndrome: An Unusually Early Presentation in a Newborn Girl**

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We report on a female newborn, the youngest patient with Zimmerman-Laband syndrome hitherto reported. She had gingival hyperplasia, bulbous soft nose and ears, hypoplastic toenails, and hyperextensibility of the joints, as well as deep palmar and plantar creases, a sign not previously described in literature.

**Key words:** gingival hyperplasia; hypermobility, joints; hyperplasia; mental retardation; nails, malformed; newborn; syndrome

Zimmermann-Laband syndrome (ZLS) is a rare disorder characterized by gingival hypertrophy, bulbous soft nose and thick, floppy ears, aplasia or dysplasia of nails and/or terminal phalanges, hyperextensibility of joints, and, in a small percentage of patients, hepatosplenomegaly, hypertrichosis, and mental retardation (1,2). This report of a female newborn is the youngest patient with ZLS so far described.

**Case Report**

Our proposita, a girl, the first child of young, nonconsanguinous parents, was admitted to the University Hospital Zagreb, Department of Pediatrics, at the age of 10 days, with suspected hypothyroidism and mucopolysaccharidosis. The pregnancy had been uneventful and the infant's birth weight was 3,600 g (75th percentile) and birth length 50 cm (50th percentile).

The examination showed characteristic formation and consistency of the nose and ears (Fig. 1). Diffuse overgrowth of the gingivae was observed, as well as of the major portion of the palatal tissue (Fig. 2); in addition, the tongue was very large. The toenails were hypoplastic. There was hyperextensibility of the metacarpophalangeal joints, and deep palmar and especially plantar creases were noted (Fig. 3). The liver was palpable 0.5 cm below the costal margin.

![Figure 1: Proband M.A. Note broad bulbous nose.](view_this_figure)

1:

![Figure 2: Proband M.A. Note bilateral swelling of rugae of broad palate and gingival hypertrophy.](view_this_figure)

3:

![Figure 3: Proband M.A. Note deep plantar creases.](view_this_figure)

Complete blood count and blood chemical tests, including thyroid function tests, and urinary amino acids and mucopolysaccharides were normal. A normal 46,XX karyotype was established. The EEG was normal. Radiological examination of the hands and feet showed aplasia of the distal phalangeal bones on both fifth toes and hypoplasia of the distal phalangeal bones on all fingers.

The mother of the proposita was normal in appearance, whereas the father (24 years old) showed some signs of ZLS, specifically gingival hypertrophy of the posterior part of the maxillary arch, and minimal soft tissue enlargement of the nose and ears.

By the father's account, he resembles his deceased mother, who allegedly has soft tissue enlargement of the nose and ears, gingival hyperplasia and deep palmar and plantar creases as well. No one in the family has liver disease, splenomegalia, or mental retardation.

**Discussion**

The total number of cases of ZLS now approaches 30. The first 11 cases belonged to two large pedigrees of East Indian ancestry, in both of which the pattern of transmission was clearly autosomal dominant (3,4). Numerous reported cases with the clinical features of ZLS occurring in other Caucasian groups have all been isolated cases (5,6). Although we have an incomplete family
pedigree, the available data point to an autosomal dominant inheritance pattern. The proposita is the youngest patient with ZLS so far described. The deep plantar creases allegedly shared with her paternal grandmother, are a clinical sign not previously described. The gingival hyperplasia central to the syndrome, and in this case already present in the newborn period, usually does not become evident until the eruption of the permanent teeth. It is anticipated that this problem will worsen as the girl grows, and that gingivectomy and gingivoplasty will be necessary. With the suggested severity of the condition in her case from its early presentation, it is probably very important for us to follow this patient closely for the earliest signs of fibrosis of the spleen and liver as well.

References

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