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BART'S SYNDROME WITH EAR MALFORMATION AND NATAL TEETH: A CASE REPORT

SUDHAKAR AJMERA[1], MUDDU GOPI KRISHNA[2], B. BALRAM[3] & AKULA AKHILA[4]

[1]Assistant Professor, Department of Pediatrics, Kakatiya Medical College and Mahatma Gandhi Memorial Hospital, Warangal, Telengana, India

[2]Junior consultant, Department of Pediatrics, Sri Sathya Sai General Hospital, Prasanthi Nilayam, Ananthapur, A.P, India

[3]Professor & Head of the Department, Department of Pediatrics, Kakatiya Medical College and Mahatma Gandhi Memorial Hospital, Warangal, Telengana, India

[4]Junior Resident, Department of Pediatrics, Kakatiya Medical College and Mahatma Gandhi Memorial Hospital, Warangal, Telangana, India

ABSTRACT

Bart's syndrome is characterized by congenital localized absence of skin, congenital epidermolysis bullosa, and associated with nail abnormalities. It is an autosomal dominant disorder with variable presentation. We report a very rare case of Bart's syndrome with multiple abnormalities. A male late preterm born to consanguineous couple, with history of similar problem in the elder sibling which died in neonatal period our case presented with absence of skin in scalp, face, trunk and extremities. Baby has deformed fingernails and eyes; ears and nose are not well formed. Peg shaped deformity of teeth, left multi cystic dysplastic kidney were noted. Baby died on the first day of life. The diagnosis of Bart's syndrome was made on clinical presentation, family history and skin biopsy.

KEYWORDS: Bart's Syndrome, Malformed Ears, Peg Shaped Natal Teeth