Images in Clinical Practice

Beckwith-Wiedemann Syndrome

A 4-hour-old male infant weighing 4.4 Kg, born at full term to a primigravida mother presented with exomphalos major (herniation of intra-abdominal contents through a wide open umbilical ring into the base of the umbilical cord) and macroglossia (Figs. *1 and 2).* Primary closure of the abdominal wall defect was done. He had a stormy post-operative period including citrobacter septicemia, staphylococcal pneumonia and neonatal hyperbilirubinemia (requiring an exchange

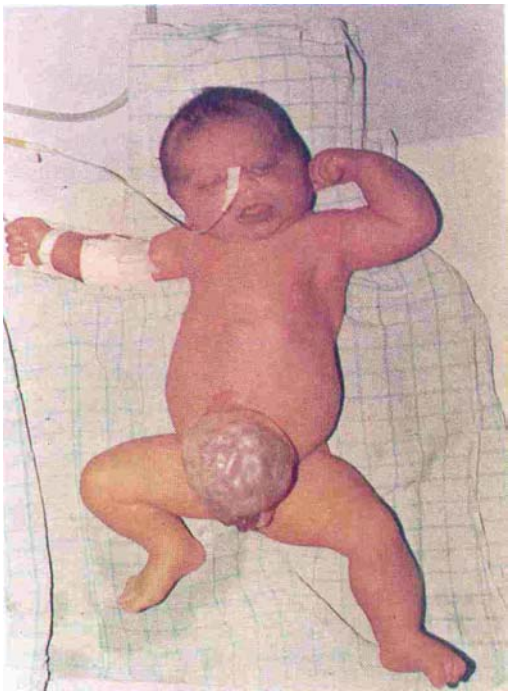

Fig. 1. Note three important features of Beckwith Wiedemann Syndrome, namely, exomphalos, gigantism, and macroglossia. Small intestine is protruding out through a small rent in the inferior aspect of exomphalos sac.

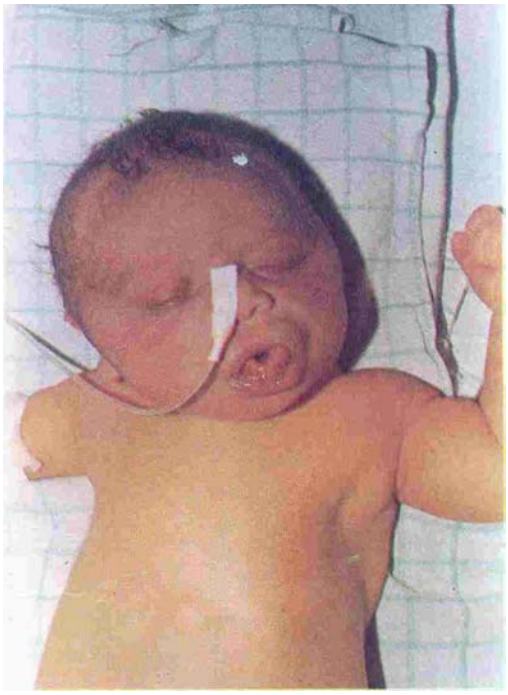

Fig. 2. Close-up of the face showing macroglossia and generally coarse features.

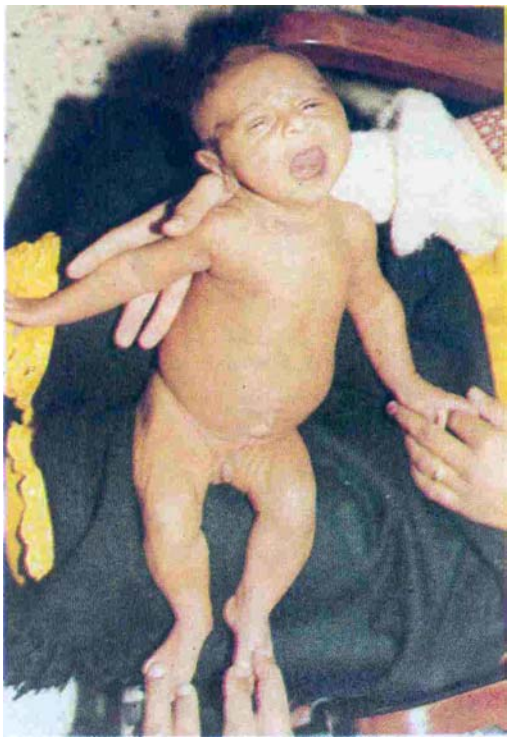

Fig. 3. Photograph of the same baby taken at 3 months of age.

transfusion). He also had intractable hypoglycemia during the first three weeks of life which was treated with 15% glucose and short-term steroid administration. During the next 2-year follow-up, he continued to have accelerated somatic growth with a disproportionately slow head growth *(Fig. 3)*. He has delayed milestones and is mentally retarded, probably related to the neonatal hypoglycemia. He has not developed hemihypertrophy or associated neoplasia.

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