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WEST AFRICAN JOURNAL OF MEDICINE



CASE REPORT AND REVIEW OF LITERATURE

Management and Outcome of a Case of Osteogenesis Imperfecta in Benue State University Teaching Hospital, Makurdi, North-Central Nigeria

Gestion et Résultats d'un cas d'Ostéogénèse Imparfaite à l'Hôpital Universitaire de l'État de Benue, Makurdi, dans le Centre-Nord du Nigeria

M. O. Ochoga[†], E. E. Eseigbe[†], A. O. Shogo^{*‡}, I. Ikuren[‡]

ABSTRACT

Osteogenesis Imperfecta is a genetic disorder of the connective tissue leading to generalised osteoporosis, fragility of the skeletal system and susceptibility to fractures of the long bones and compression of the vertebrae from mild or inconsequential trauma. It is one of the rare diseases known to mankind. It has no definitive cure and treatment is essentially supportive. We present below a preterm male neonate who was seen 5-hours after birth with abnormal posturing, abnormal shape of the head and limbs. There was a positive family history of delivery of a baby with similar history and outcome. A skeletal survey revealed multiple fractures involving the clavicle, humerus, femur, tibia and fibula. He was managed as a case of Osteogenesis Imperfecta. This was the first case out of 1,445 newborns admitted into the Unit. He was discharged after 2-weeks of hospitalisation but died at 6-weeks of age following progressively worsening episodes of respiratory distress. **WAJM 2021; 38(1): 80–83.**

Keywords: Osteogenesis imperfecta, congenital fractures, Benue, Nigeria.

RÉSUMÉ

L'ostéogénèse imparfaite est une maladie génétique du tissu conjonctif qui entraîne une ostéoporose généralisée, une fragilité du système squelettique et une prédisposition aux fractures des os longs et à la compression des vertèbres à la suite d'un traumatisme léger ou sans conséquence. Il s'agit d'une des maladies rares connues de l'humanité. Elle n'a pas de remède définitif et le traitement est essentiellement de soutien. Nous présentons ci-dessous un nouveau-né mâle prématuré qui a été vu 5 heures après la naissance avec une posture anormale, une forme anormale de la tête et des membres. Il y avait des antécédents familiaux positifs d'accouchement d'un bébé avec des antécédents et des résultats similaires. Une étude du squelette a révélé de multiples fractures impliquant la clavicule, l'humérus, le fémur, le tibia et le péroné. Il a été traité comme un cas d'ostéogénèse imparfaite. C'était le premier cas sur les 1445 nouveau-nés admis dans l'unité. Il est sorti après deux semaines d'hospitalisation mais est mort à l'âge de six semaines à la suite d'épisodes de détresse respiratoire qui se sont progressivement aggravés. **WAJM 2021; 38(1): 80–83.**

Mots-clés: Osteogenesis imperfecta, fractures congénitales, Bénoué, Nigeria.

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Abbreviations: