Abstract # 231

**Anaesthetic Management of a Primigravida Patient with Cleidocranial Dysplasia Undergoing Caesarean Section**

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A 21 year old G1P0 patient with cleidocranial dysplasia required an urgent caesarean section at 39/40 for breech presentation in a previously uneventful pregnancy. Past history revealed numerous dental procedures, congenital absence of clavicles, persistent fontanelles and a benign heart murmur.

Airway assessment revealed supernumerary teeth but no features predictive of difficult intubation. Cardiorespiratory assessment revealed obvious narrowing of the upper thorax and absence of clavicles. Assessment of her spine revealed normal curvatures and thus a subarachnoid block was performed to provide surgical anaesthesia.

Delivery and recovery were uneventful and the patient was subsequently discharged home.

Cleidocranial dysplasia is a rare congenital disorder with equal sex distribution characterised by absent or hypoplastic clavicles, multiple supernumerary teeth and open sagittal sutures and fontanelles. It is commonly due to a mutation which affects osteoblast differentiation.

Craniofacial abnormalities include a high arched palate, mandibular prognathism, maxillary underdevelopment and multiple impacted and supernumerary teeth which may affect airway management.

The thorax may be narrow and cone shaped with absent or hypoplastic clavicles, scapulae and ribs that may predispose to respiratory failure during infancy or in the post-operative period. The absence of landmarks may result in difficulties in obtaining central venous access in the neck necessitating ultrasound identification.

Vertebral abnormalities include kyphosis, scoliosis and spina bifida occulta which may make positioning and epidural insertion technically difficult and subarachnoid block unpredictable.

Pelvic abnormalities include a contracted pelvis and may necessitate delivery via caesarean section. Other skeletal abnormalities involve the limbs such as hypermobility and dislocation which emphasises the importance of careful positioning to prevent injury.

Non-skeletal manifestations include recurrent respiratory tract infection, conductive deafness and syringomyelia.

There are few articles published to date which describe the anaesthetic management of these patients and awareness of the skeletal and extra-skeletal manifestations can prepare the anaesthetist to the challenges.

**References:**