

A Rare Entity: Case Report on Anaesthetic Management in Robinow Syndrome

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Abstract : A five-year-old male child born from non-consanguineous marriage, who presented with complaints of growth retardation and no appreciable increase in the penile size since birth and he was posted for de-gloving of penis with dissection of corpora under anaesthesia. After thorough preoperative evaluation it was revealed that patient had peculiar facial dysmorphism that of Robinow Syndrome, high arched palate, Mallampati grade III, mesomelic limbs, scoliotic spine and short stature. All routine investigation were within normal limit, electrocardiography (ECG) and 2D-Echocardiography (ECHO) were normal. In antero-posterior roentgenogram chest showed butterfly and hemivertebrae at multiple levels. The patient was considered to be ASA II. On the day of surgery after ensuring fasting of 6 hours, patient was taken in operation theatre, all standard ASA monitoring was done with ECG, non-invasive blood pressure, peripheral oxygen saturation (SpO₂) and body temperature. The patient was pre-oxygenated with 100% oxygen with anatomical face mask. General anaesthesia was induced with Sevoflurane 1-8%, and airway was secured with an appropriate size supraglottic airway and anaesthesia was maintained with nitrous oxide and oxygen in 1:1 ratio along with sevoflurane 2%. An ultrasound guided caudal block was given owing to the skeletal deformities making it difficult even under USG guidance. Post operatively patient was given supportive care with proper hydration, antibiotics, anti-inflammatory and analgesics. He was discharged the next day and followed up weekly for a month. **DISCUSSION** Robinow syndrome is genetically inherited as autosomal dominant, autosomal recessive or heterogenous disorder involving tyrosine kinase ROR2 gene located on chromosome 9. It has low incidence with no preponderance for any gender. Though intelligence is normal but developmental delay and mental retardation occurs in 20%cases

Keywords : Robinow Syndrome, dwarfism, paediatric, anaesthesia

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