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AN ANAESTHETIC CHALLENGE: MANAGEMENT OF ASYMMETRIC HYPOCHONDRIAC DWARF

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ABSTRACT

: Dwarfism is restricted growth due to different disorders, where height varies between 122 to 147 cm. Hypochondriac dwarfism a form of achondroplasia, transmitted as an autosomal dominant trait, presents with difficult airway, spinal deformities, cardiac and neurological involvement, disturbed calcium metabolism, thermal regulation and coagulation.[1] with social and psychological problems promoting drug abuse . Achondroplasia has been known for many years but hypochondria was first reported in English literature by Beals (1969). The features are similar in both but less severe in hypochondria and does not involve skull.[2]. Anaesthetic management is a challenge for abnormal physical and physiological presentation with lack of guidelines regarding calculation of drug doses in accordance to disproportion in their height, weight and age.

Key Words: Hypochondriac dwarf, spinal deformities , difficult airway , neuraxial block, drug doses, endotracheal tube size.

INTRODUCTION

Case Report: A 122cm , 25 Kg , 20 yrs hypochondriac male dwarf, working in fields irregularly , reported for ureteroscopic removal of left ureterovesical junction calculus . He

had pectus carinatum, kyphoscoliosis, bilateral genu valgum , narrow intervertebral disc spaces, poorly developed and hypotonic skeletal muscles and normally developed external genitalia. Obstructive sleep apnoea and snoring history was negative. He did not report any comorbidity, medical treatment or drug abuse. No exposure to anaesthesia and surgery was found.

On examination he had normal sensorium and exaggerated knee jerks. The vertical distance between inferior angle of scapula to iliac crest was 5.5cm in erect posture confirming narrow intervertebral disc spaces and possibility of spinal canal stenosis .

Head, chest and abdominal circumferences were 51, 70 and 63cm respectively.

Airway revealed short neck, protruding mandible, malocclusion, high arched palate, large tongue and adenoids with excessive oronasal secretions. Neck movements, mouth opening, bilateral air entry and breath holding time recorded were normal. . Mallampatti grade was II . Clinically congenital cardiac anomalies were not present. Apex beat found in third intercostal space. ECG showed right bundle branch block, premature atrial complexes and left axis deviation.

Investigation reports showed Hb11.7gm% ,serum calcium 8.2 mg/dl and normal vitamin D3. Total and differential leucocyte count, random blood sugar, liver and renal function test, platelet count, bleeding and clotting time, X-ray chest and 2D echo study were within acceptable limits with non reactive viral markers.

Anaesthetic procedure: Informed written consent obtained for general anaesthesia. Base line haemodynamic parameters recorded and found within normal limits. Following start of ringer lactate infusion with 20G intracath in supine position, preoxygenation with 100% oxygen and premedication with glycopyrrolate 0.2mg, ranitidine 50mg, ondenseteron 4mg, fentanyl 50µgm, midazolam 0.5mg IV was done. Intravenous induction of anaesthesia with propofol 2.5mg/kg done with no difficulty in mask ventilation. A direct laryngoscopy was performed to asses, largest diameter of endotracheal tube, any possibility of difficult intubation and to exclude laryngeal hypoplasia.

Relaxation for intubation with vecuronium bromide 0.1 mg,/kg was given.Above doses could not provide adequacy for intubation so two additional supplementary doses of propofol 20mg + 20mg and 0.5mg of vecuronium over the calculated dose according to body weight of dwarf were needed. Patient was oxygenated by non invasive intermittent positive pressure ventilation for 3minutes. 7 mm ID cuffed endotracheal tube was passed by advancing it

gently and slowly step by step to avoid tracheal injury due to predictable shortening of tracheal length and tracheomalacia. Position of endotracheal tube was confirmed by auscultation and fixation done at 16cm. Anaesthesia maintained by N2O + O2 1:1 and isoflurane 0.6 to 1.5 MAC on volume controlled ventilatory support to maintain normocarbia and 100% oxygen saturation. 10 minutes later an episode of tachycardia and rise in blood pressure was recorded and bolus dose of propofol 20mg was given to overcome this. Surgery continued for 75 minutes. Injection xylocard 2%, 2ml to overcome sympathetic stimulation during extubation resulted in smooth reversal by neostigmine 2.5mg +glycopyrrolate 0.5mg, intravenous slowly.

Ringer lactate 750 ml and normal saline 350ml was infused intra operatively. His pulse, NIBP, SpO2, ECG and ETCo2 were monitored continuously and found to be stable within normal range. For post operative analgesia paracetamol 1gm infusion was given. His post operative stay was uneventful with discharge on 7th day.

Discussion: Anaesthetic management both neuraxial and general of hypochondriac and achondroplastic dwarfs imposes many specific difficulties as they have multiple problems involving airway, pulmonary, cardiac & neurological systems along with disturbances of thermal regulation and coagulation. Disturbances of calcium metabolism and bone formation make them prone to fractures.[3]

To choose the modality for anaesthesia management for them an anaesthetist has to asses predictable and unforeseen difficulties and complications of general anaesthesia and neuraxial blockade.

For the selection of anaesthetic plan in our case we considered subarachanoid block and epidural anaesthesia but found it to be difficult technically as well as for calculation of drug dose which could provide desired effect without giving rise to any complication like high spinal or incomplete blockade. No definite direction regarding calculation of drug doses is found in literature for such cases. Accurate blood pressure measurement during spinal and epidural anaesthesia makes it essential to use an appropriate size of blood pressure measuring cuff covering 2/3rd of the upper arm which again is difficult to obtain for dwarfs. Specially in cases of osteogenesis imperfecta found in hypochondriac dwarfs this problem can lead to pathological fractures. Even then several successfully performed epidural and spinal anaesthesia techniques for caesarean section in different types of dwarfism are reported.[4,5,11,12,13]

Difficulties related to general anaesthesia are encountered due to difficult airways, large tongue, large tonsils, stiffness and immobility of temporomandibular joint, narrow nasal passages due to mucopolysaccharide deposition ,[3] large adenoids, unpredictable shortening of diameter and length of trachea along with large amount of nasal secretions. These are exaggerated by cervical abnormalities, sternal prominences associated with pectus carinatum as was found in our patient. Occasional abnormalities of the base of skull may limit neck extension which interferes with laryngoscopy[1,3]. There could be restrictive or obstructive lung abnormalities along with congenital or acquired cardiovascular abnormalities which may create additional problems. A positive history of obstructive sleep apnoea should be taken very seriously as such patients may show signs of upper airway obstruction after sedation or induction of anaesthesia.[1,3] One should be prepared with difficult airway management cart to immediately switch over to other techniques of intubation like fiber optic intubation and emergency tracheostomy for which we were prepared. In our patient we did not anticipate much difficulties during general anaesthesia as patient was in Mallampatti grade 2 but the presence of large tongue , high arched palate, large adenoids, large amount of secretions, pectus carinatum and indefinite diameter and length of trachea alerted us to face the situation of difficult airway. Pectus carinatum could produce restrictive respiratory pattern and pose intra operative difficulty in mechanical ventilation with co-existing obstructive lung disease.

Another difficulty arises in calculating drug doses, judging the size (internal diameter)and length of endotracheal tube at which fixation should be done after intubation. This is difficult because of discrepancy between age, height and body weight of dwarfs. In absence of any guide line for above we used indirect formula (Weech's formula) which was to know the approximate age at which a male child attains a height of 122 cm.[6]

(Age x 6) + 77 = height in cm

(Age x 6) + 77 = 122 cm; age = 7.5 yrs

According to Paul[7] normal male at 7.5yrs, weighs 24 kg which was nearly the weight of our patient

(25 kg)so we calculated the anaesthetic drug doses accordingly.

Formula used[8] for tube length in cm was (age/2)+12 =(7.5/2)+12=15.75cm = 16cm approx.

To know the internal diameter of the endotracheal tube we performed direct laryngoscopy and found 7mm size appropriate for the case. This selection of size and length is important because according to Hagens Poiseuille equation:

Flow of gases $\propto \pi R4\Delta P/(8\eta L)$

Where R & L are radius and length of ETT, η viscosity of flowing gas.

In the presence of restrictive respiratory pattern and probable occurrence of obstructive pattern largest size internal diameter tube was chosen because flow of gases are proportionate to the fourth power of radius

Laryngeal hypoplasia may require smaller internal diameter of endotracheal tube than predicted. Awake intubation or inhalational induction with oxygen and halothane is recommended. Muscle relaxants should be avoided until positive pressure ventilation by mask can be ensured[3]. In our patient we preferred to use vecuronium and cautiously avoided succinylcholine due to chances of dysrrhythmias, bradycardia and hyperkalemia which could occur in our patient who had pre existing ECG changes .

Muscular twitchings caused by succinylcholine can produce fractures in cases of osteogenesis imperfecta found in hypochondroplsia. [3,9]

Neck movements during laryngoscopy and intubation may result in atlanto occipital dislocation because of laxicity of these ligaments which may cause sudden death. Subglottic stenosis, tracheo esophageal fistula and chronic pulmonary infection may give rise to various post operative respiratory complications.

Surgeon used irrigating fluid glycine and surgery lasted for more than one hour so we added normal saline to cope up for possibility of hyponatremia towards end of surgery [1].

These patients can develop hyperthermia and should be treated by external cooling. Coagulation dysfunction can lead to haemostatic problems.[3] Pre operative evaluation in view of this problem was done in our case. Fresh frozen plasma and platelates were made available although patient did not require them.

CONCLUSION

We can get good results in challenging anaesthetic management of hypochondriac dwarfs by proper pre anaesthetic evaluations in view of abnormalities and complications involving various systems, sleep apnoea, airway, selection of anaesthetic drugs their doses, type of surgery proposed and socio psychological problems resulting into drug abuse. Suitability of anaesthetic plans should be evaluated for each dwarf individually. As was observed in our patient drug doses should be considered according to an average built adult of that age rather than body weight of the dwarf since additional doses were required intraoperatively .[10] ET tube size chosen should be largest detected by direct laryngoscopy. Cumulative effect of above considered points are supposed factors for successful outcome.

We wish to share experiences of other expert anaesthesiologists regarding management of such cases.

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