AS but asymptomatic and uneventful antenatal course.

**CASE REPORT:** A 25 year old 34 week G1P0 achondroplastic female with severe AS was admitted to the hospital for elective caesarean section. An elective surgery was planned, on pre anaesthetic check-up she has papers suggestive of diagnosed achondroplasia and severe AS. She was currently on medication Carvedilol 3.1 mg BD and Toresamide 5mg OD.

The physical examination demonstrated an amicable patient who was 114 cm tall and weighed 34 kgs. On GPE she had a normal size face, short neck, depressed AP diameter of chest and distended abdomen. On cardiac auscultation, she was diagnosed to have crescendo- decrescendo systolic ejection murmur. ECG was suggestive of LV hypertrophy with sinus rhythm, short PR and V4-V6 ST segment abnormality. Her echo showed severe AS with mild MR with aortic root diameter 14 mm, EF=60%, AVPG mean/ AVPG max = 55/100. Respiratory system examination showed symmetrical barrel shaped chest with AP diameter almost equal to transverse diameter. RR and rhythm were normal. On breathing chest expansion was 1-2 cm.

Palpation showed trachea in midline and on auscultation decreased breath sounds with bilateral and normal vesicular breathing but prolong expiration with expiratory ronchi was heard. PFT was suggestive of early small airway obstruction and severe airway obstruction. BHT was 15 seconds with NYHA grading 3.

Airway examination showed adequate mouth opening, MPG 2, no M/A/L dentition and neck movements were adequate. Spine examination showed central vertebrae and appropriate spinal curves but space between vertebrae was not appreciated on palpation.

Her routine blood investigations were with in normal limits. This parturient had several comorbidities so this was supposed to be a planned elective LSCS and was under evaluation. Certain investigations like MRI neck, cardiac surgeon opinion and ENT opinion for expected difficult airway were due and to be done on coming working day.

But unfortunately patient landed up in an emergency with fetal distress due to meconium stained liquor. So she was taken for emergency LSCS with high risk consent and proper explanation of procedure and post operative risks of anaesthesia. On table PAC was done. Her vitals were, BP-110/70 mm of hg, HR- 82 bpm and spo2- 100% on room air. ECG was same as before. Right sided pelvic wedge was applied and two 18 gauge cannula were secured. Injection Ranitidine 50 mg with injMetoclopromide 10 mg, injloxicard 50mg and inj midazolam 2mg iv slow were given as premedication.

Intra-arterial cannulation was done in left radial artery in favour of invasive BP monitoring. Patient was preoxygenated for 5 minutes with 100% oxygen and was induced with inj Fentanyl 60 mcg+ injEtomidate 14 mg and injscholine 60 mg for RSI. Direct laryngoscopy showed CL grading 2a. An attempt of intubation was taken with cuffed ETT size 6.5 but could not be inserted then bag and mask was resumed and after some time 2$^{nd}$ attempt was taken with ETT size 6.0, which also failed. Immediately senior anaesthesiologist took the charge, bag and mask and mask was done followed by another attempt with ETT size 5.5 which just

reached upto vocal cords but could not proceed further. Bains circuit was connected to this tube and a positive chest rise and adequate bilateral air entry was noted on auscultation and confirmed with ETCO2.

We failed to advance the tube further but succeeded to ventilate the patient. Frequent oropharyngeal suction was done in between the multiple attempts. Incision was given and case was conducted with tube held with hand tightly just at the angle of mouth and ventilation was assisted with bains with high pressure and respiratory rate. Intra operative maintenance was done with inhalational isoflurane 0.5 MAC and inj fentanyl 30 mcg in divided doses. Sympathetic stress response was suppressed with injesmolol. Hartman's iv fluid was given with 20ml/kg. injparacetamol 300mg was given for pain management.

Baby was delivered and inj. oxytocin 5 IU was given. On birth baby cried with APGAR score 8. After completion of case tube was removed when patient became conscious, oriented and with adequate muscle power and airway reflexes. We requested obstetrician to continue same treatment and monitor postoperatively and her subsequent course in hospital was uneventful. Later she was referred to cardiac surgery department.

Table1:-showing patient parameters

| Introp monitoring | SBP(mm of hg) | DBP (mm of hg) | HR (bpm) | SPO2(%) |
|---|---|---|---|---|
| baseline | 110 | 70 | 82 | 100 |
| induction | 99 | 65 | 72 | 100 |
| intubation | 135 | 69 | 111 | 100 |
| 2 min | 140 | 77 | 135 | 99 |
| 5 min | 139 | 79 | 140 | 99 |
| 10 min | 120 | 80 | 103 | 99 |
| 15 min | 114 | 74 | 99 | 100 |
| 20 min | 122 | 75 | 91 | 100 |
| postop | 128 | 64 | 82 | 100 |

**DISCUSSION:**

There are different types of dwarfism, many of which have specific implications for anaesthesiologist. Although being an anaesthetist we encounter such patients.(2) Females are more affected than males. Anaesthesia whether general or regional poses many potential problems in these patients specifically during later stages of pregnancy. These dwarfs have low fertility rate but those who do conceive, often require a lscs due to cardiac compromise, ceohalopelvic disproportion.(3)

These patients have several derangements which have potential for complications during administration of anaesthesia. Narrow nasal passages and nasopharynx, large tongue and mandible,short neck,craniofacial abnormalities such as macrocephaly, frontal bossing, depressed nasal bridge, maxillary hypoplasia and occipitalisation of the first cervical vertebra can all lead to difficulty in mask ventilation ,glottic opening visualisation and intubation(4) .Tracheomalacia may also be present .Review of all previous anaesthetic records regarding airway management should be done. Preoperative evaluation of the airway by various radiological investigations like CT scan and MRI is indicated.Awake fiberoptic intubation has been recommended (1)

Restrictive lung disease is common in these patients due to thoracic dysplasia(rib hypoplasia), thoracic lordosis and thoracic kyphoscoliosis .There is ventilation perfusion mismatching due to decreased FRC and increased closing volume promoting atelectasis which require thorough evaluation of pulmonary function. In addition to this there is a tendency for sleep apnea which may be central or obstructive in nature due to craniofacial abnormalities and hypotonia of

upper airway muscles(2) .These can lead to development of pulmonary hypertension.Compression of spinal nerve roots can lead to development of neuromuscular weakness. Because of all these problems there may be difficulty in maintaining oxygenation during general anaesthesia and post operative mechanical ventilation may be required. Spinal cord and nerve root compression syndrome due to prolapse of intervertebral discs into the stenosed spinal canal can lead to slowly progressing Para paresis, quadriparesis, sensory deficits, sphincter deficits, autonomic hyperreflexia(2) . Suxamethonium should be avoided as it can lead to life threatening hyperkalaemia in presence of peripheral denervation There is constriction of the spinal canal resulting in narrowing of subarachnoid and epidural spaces which may cause difficulty in establishment of central neuraxial blockade. Pregnancy in achondroplastic dwarfs leads to further problems as the uterus remains an entirely intraabdominal organ because there is failure of normal sized fetal head to engage in narrow pelvic inlet(3) . Thus, diaphragmatic splinting occurs causing further reduction in FRC and there is severe aortocaval compression. There are several reports of parturient with achondroplasia undergoing caesarean section successfully under both general anaesthesia(4-6)as well as under central neuraxial blockade however difficult intubation has been encountered(7,8) The possibility of atlanto-axial instability, restrictive lung disease and neuromuscular weakness would influence the decision to avoid general anaesthesia. In our patient we preferred giving general anaesthesia it was an emergency for life of baby due to MSL. The risk of general anaesthesia versus regional anaesthesia must be considered for each patient as severity of spinal abnormality can vary considerably.The chances of successful epidural anaesthesia in achondroplastic patient will be maximized by careful patient selection and preparation, with discussion of risks and possibility of technical complications , optimal positioning and cautious dose titration.

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