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Anaesthesiology

ANAESTHETIC MANAGEMENTIN MUCOPOLYSACCHARIDOSIS

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ABSTRACT The high prevalence of airway obstruction and cardiovascular manifestation possess high anaesthtic risk in morquio's dieases. which can be minimized by proper preop assessment, check ventilation before giving muscle relaxen tand preparing difficult airway cart before intubation.

Aim: Airway & anaesthetic management in Mucopolysaccharidosis type IV.

KEYWORDS : Laryngoscopy, difficult airway cart(bougie,airway,uncuffed and cuffed smaller sized et tubes)

Aim:
Airway & anaestheticmanagement in Mucopolysaccharidosis type IV.

Introduction:
Mucopolysaccharidosisrepresents a group of rarelysosomestorage disorders associated with accumulation of glycosaminoglycans in tissuesand organs.

Thehigh prevalence ofairway obstruction and cardiovascular manifestations possess highanaestheticrisk.

Case:
A 16year old male child having congenitalhydrocephalus operated for VP shunt.Patient isdiagnosed as mucopolysaccaridosis "Morquio's disease "in 2008 andpresentedwith decrease vision inboth eyes since2 years and posted for Vpshunt on administration of study drug, after induction, at laryngoscopy and intotubation, at 1,2,3,5, and 10 mins after intubation.

Patient had height 118cm, weight 16kg,head circumference 51 cm, large tongue,short neck, highanterior larynx, proptosis,cervical instability& skeletaldeformity.

X-ray
ychest showed trachea shiftedto rightside.

X-raydorsolumbar spine showedposterior displacement of L2vertebraresulting in kyphotic deformity

2D echo showed Moderate MS, ModerateMR & Bicuspid aorticvalve.

During Airway examination we found Mallampati grade IIIwith adequate Mouth opening & 5cm ThyromentalDistance.

Difficultairway cart was kept ready.

Anaesthetic management:
On the day of surgery, high risk informed consentwas taken & vitals were monitored.

Preoperatively nebulizationwas done. Premedication was given. Pulse 149/min, NIBP 114/76mmhg, SPO2 99%

Preoxygenation:100% oxygen given bybains circuit at 6-8L/min

Induction: Inj. Propofol 2.5mg/kg i.v.,ventilation test was done. Asventilation was possible, we had givenInj.Scoline 2mg/kg i.v

Laryngoscopywas done with **McCoy blade** and ETtube no.5.5 cuffed inserted. Bilateral air entry checkedand tube fixed

Maintenance of anaesthesia: O2 and sevoflurane
Inj. Atracurium - 0.5mg/kg loadingi.v.,
0.1mg/kg incremental i.v.

Intraoperative vitals were monitored.

Atthe end of surgery,reversal inj. Neostigmine0.05mg/kg wasgiven.

Afterachieving consciousness & muscle tones ofpatient ET tube wasremoved with tube exchanger. Postoperatively period wasuneventful.

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