ANAESTHETIC MANAGEMENT FOR PATIENT OF LENS DISLOCATION WITH MARFAN SYNDROME

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ABSTRACT

INTRODUCTION: Marfan syndrome is an autosomal dominant, multisystem disease with a reported incidence of 1 in 3000 to 5000 individuals. Classic manifestations involve ocular (lens dislocation, myopia), cardiovascular (aortic root dilatation with aortic regurgitation, mitral valve prolapse with mitral regurgitation), and musculoskeletal abnormalities (long bone overgrowth, scoliosis, kyphosis, joint hypermobility).

CASE SUMMARY: A 20-year-old female, 47 kg with a history of Marfan syndrome presented to the comprehensive ophthalmology SMEC hospital reporting a progressive decrease in vision and worsening glare in both eyes during the last few months. She has been examined by ophthalmologists and was told that her crystalline lenses were subluxated in both eyes. Examination of the oral cavity revealed a high arched palate, grade II mallampati coupled with pointy chin indicating possible intubation problem. Chest x-ray showed scoliosis thoracalis dextra. Other examinations were within normal limits. Patient was preoxygenated, premedication given include injection of Fentanyl, Midazolam and lidocain. Patient was induced with Propofol IV to induce deep sleep. After epiglottis could be visualized atracurium was injected, no. 7 cuffed endotracheal tube was passed through right mouth tip and the tip was manipulated below the epiglottis to pass in the larynx smoothly to prevent the increase of intraocular pressure. Anaesthesia was maintained by Oxygen, Air, sevoflurane. Intraoperatively, patient hemodynamic remain stable with minimal blood loss. Surgery lasted for 1 hour 15 minutes and in the end patient was extubated in operating room. Patient transferred to recovery room and hemodynamic was closely monitored.

CONCLUSION: General anesthesia with endotracheal tube is the chosen technique for this patient. It is prudent to avoid excessive elevation of intraocular pressure as can occur during laryngoscopy, intubation, or response to painful surgical stimulation. Thus, intubation should be performed gently and smoothly with deep extubation.