



Case Report

Anesthetic Challenges of Cataract Surgery in a Rare Case of Hutchinson Gilford Progeria Syndrome

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ABSTRACT

Hutchinson Gilford Progeria Syndrome (HGPS) is a rare genetic disease characterized by accelerated ageing with severe growth retardation. This syndrome involves multiple organ systems like cardiovascular system, excretory system and central nervous system. Due to abnormal dentition, difficult airway and ischemic heart disease it is real challenge for anaesthesiologist. It is clubbing of pediatric and geriatric anaesthesia. Very few cases of general anaesthesia management in developmental cataract surgery were reported in literature. We report a case of successful general anaesthesia management of developmental cataract excision of case of Hutchinson Gilford Progeria Syndrome (HGPS).

Keywords: Hutchinson Gilford Progeria Syndrome (HGPS), developmental cataract, general anaesthesia.

INTRODUCTION

HGPS (Hutchinson Gilford Progeria syndrome) is a rare genetic disorder caused by de novo point mutations in LMNA gene. The incidence is in 1 in 8 million. ^[1] In the literature very few cases reported of general anaesthesia management of children with this syndrome. Herein, we report a case of anesthetic management in developmental cataract excision in 11-year-old progeria child.

CASE REPORT

An 11-year-old girl presented with dimness of vision of left eye since 6 months and diagnosed as developmental cataract. Her weight was only 17 kg. During pre-

anaesthetic evaluation due to facial features poor dentitions, scanty hairs and wrinkled dry skin. Patient was diagnosed as Progeria. (Figure 1)



Figure 1. It shows 11 yr old girl with adult facial features.

She was full term normal vaginal delivery. Her milestones were normally attained and no mental retardation noted. She was immunized properly till date. On general examination vital parameters were within normal limits. PR- 70/mins with occasional ectopic beats, BP- 100/60 mm of Hg and RR- 18 beats/min. Her neurological, respiratory and cardiovascular examination were normal.

Her mouth opening was MPC grade II. Incisors were widely spaced. Neck movements were normal. Routine blood investigations were within normal limits.

ECG was showing right axis deviation with occasional premature atrial beat (3-4/min). 2 D Echocardiography was not done due to unavailability of cardiologist.

ECG monitor, pulse oxymeter and NIBP monitor applied. Peripheral intravenous canula no 22 inserted and Isolyte -P started. Baby premedicated with Inj. Glycopyrolate 0.08mg IV and Inj. Midazolam 0.5mg IV. Difficult intubation preparation made. Induction accomplished with Inj. Thiopentone sodium 100mg IV and intubated under effect of Inj. Suxamethonium 35mg IV using cuffed PVC tube no 5.5 with aid of external laryngeal manipulation. Anaesthesia maintained on oxygen, nitrous oxide and halothane. Muscle relaxation achieved with Inj. Vecuronium 1.5mg IV.

Peribulbar block given using inj. Bupivacaine 0.5% 5cc for postoperative analgesia and to reduce intraocular pressure. Surgery was uneventful and anaesthesia was reversed with Inj. Neostigmine 0.8mg + Glycopyrrolate 0.15 mg IV. Patient extubated uneventfully and observed for 24 hrs. She is discharged from hospital on 3rd post operative day.

DISCUSSION

Hutchinson Gilford Syndrome (Progeria, Premature aging) is very rare genetic disorder. ^[1] This is Autosomal recessive disorder that usually becomes clinically apparent after 6 months. This condition firstly described by Hutchinson in 1886 and was termed as Progeria by Gilford in 1901. ^[1] Defective Lamina-A proteins due to mutations in LMNA gene on chromosome L causes cellular instability and leads to premature aging. It is more prevalent in males. ^[1,2] Effective treatment is not yet available. These patients died at the age of 13 years and most common cause is ischemic heart disease and stroke. ^[2] These patients often require surgeries because they are vulnerable for pathological fractures and dislocations. Very few cases reported of general anaesthesia management of these patients. Anaesthesia management is challenging due to potential difficult airway, advanced cardiovascular disease and risk of perioperative hypothermia and hypoglycemia. ^[3-8] These babies are short statured with stiff and predominant joints. Skin is dry and wrinkled. Craniofacial and dental manifestations include mandibular and maxillary hypoplasia, dental inelasticity. ^[8,9] Repeated failed intubation attempt even with fiberoptic bronchoscope reported. ^[3-5] Propofol infusion syndrome is reported in one case. ^[10] In spite of these changes they are emotionally and intellectually children only so they should be treated according to age.

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