

CASE REPORT

Anesthesia Management in Rare Case: Cutis Laxa Syndrome

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Cutis laxa is rare and heterogeneous group of disorders related to abnormalities in elastic tissue. It may be autosomal recessive, autosomal dominant, X linked or acquired. Acquired cutis laxa has developed after a febrile illness, inflammatory skin disease such a lupus erythematosus or erythema multiform, amyloidosis, hypersensitivity reaction to penicillin and in infants born from women who were taking penicillamine. Patients with cutis laxa have facial features, pulmonary emphysema, cor-pulmonale, and right-sided heart failure. We present a case with cutis laxa undescended testis, and mild pulmonary stenosis.

Keywords: cutis laxa; pulmonary stenosis; loose skin

A 21 months old male boy weighing 12 Kg was presented to our hospital for bilateral orchiopexy. On the preoperative anesthesia visit, he was mentally sick, and has loose and thin skin. Sagging jaws, hooked nose with everted nostrils, short columella, long upper lip, everted lower eyelids (Figure 1). He was afebrile and pink without respiratory distress and edema and ascites. Pulse rate was 125/minute, regular and with sinus rhythm and blood pressure was 100/50 mm Hg. Chest was in normal. Respiratory rate was 24/minute. Auscultation of chest was normal both sides. Heart sounds were normal. Airway examination revealed a Mallampati Class III, and normal range of movements at atlanto-occipital joint. Laboratory test showed WBC (white Blood cell), 7.92 X 10³ /uL, RBC (Red Blood cell) 4.51X 10³/uL and platelets 262 X 10³ /uL. Hemoglobin level was 11.5 g/dl with a hematocrit of 33.8%. Prothrombin time was 13 seconds, Partial thromboplastin time was 30 seconds and INR was 1. Blood sugar and renal function tests were normal preoperatively. In LFT (Liver function test), AST was 138 and ALT was 43. Chest x ray showed mild hyperinflation of both the lungs and normal shadow of heart. ECG (Electrocardiography) was normal and echocardiography showed a small PFO and mild pulmonary stenosis. Abdominal sonography was normal, however, cranial sonography showed mildly dilated lateral ventricle and mild hydrocephalus. This patient is the first child in his family and with healthy parents. In past medical history, he was born at 35 weeks gestational age, because the mother was

hypertensive and as such was admitted for 10 days in the Neonatal ICU (NICU) after birth. His birth weight was 2.8 Kg. In addition to laxity of skin, he had bilateral undescended testes. On the basis of clinical examination and skin biopsy just after his birth he was labeled as a case of congenital Cutis Laxa. During his early infancy, he had history of hospitalization because of acute bronchopneumonia. During his life several admissions for pneumonia and asthma occurred. A week before surgery, salbutamol spray 2 puffs were started and on the day of surgery hydrocortisone 5 mg /kg was given.

On the day of surgery, 30 minutes before entrance to operating room patient received 5 mg/Kg ketamine and 0.5 mg/Kg midazolam orally. After sedation and anesthetizing the skin with cutaneous lidocaine an i.v. cannula was inserted. Pulse oximetry probe, ECG, and non-invasive Blood pressure monitors were applied. As difficult intubation was anticipated, fiberoptic and tracheostomy sets were made ready and, induction started with thiopental sodium 7 mg/Kg and anesthesia followed by spontaneous breathing in sevoflurane 5% in oxygen 100%, then fentanyl 1 mic/Kg injected. After appropriate depth of anesthesia, laryngoscopy was done, grade III Cormack Lehane was seen and after external pressure by second person and by help of respiratory sound patient was intubated with ETT size 4.5 mm. SUPA without cuff. After intubation, endotracheal tube was checked with End tidal CO₂ monitor and with auscultation of both lungs, and it was fixed and mechanical ventilation started and facilitated with injection of atracurium 5 mg intravenously. Anesthesia was maintained with O₂ and Isoflurane 1.5%. Normal saline solution was injected 50 ml/hour. The procedure took about 90 minutes. During the procedure there wasn't any problem in hemodynamics and airway pressure. Atropine 0.025 mg and neostigmine 0.5 mg were given to antagonize the residual effects of atracurium. Trachea was extubated in lateral position and then he was sent to the recovery room, after 30 minutes in recovery and stability and normal lung sounds, patient was transferred to surgical ward.

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Figure 1



Sagging jowl hooked nose with everted nostrils, short columella, long upper lip, and prominent low set ears

Discussion

Patients with cutis laxa have facial features, aged appearance with sagging jaws, hooked nose with everted nostrils, short columella, long upper lip, everted lower eyelids [1-2], pulmonary emphysema, cor-pulmonale, and right-sided heart failure caused by pulmonary disease have been commonly described. Pulmonary artery stenosis has been occasionally described in the literature in association with AR CL [3-4]. Cutis Laxa is a rare disorder of elastic tissue characterized by loose, inelastic skin that hangs in folds [5] is inherited or acquired [1]. Presentation of this disorder and mode of inheritance shows considerable heterogeneity. Primary or congenital Cutis Laxa is from birth and depends on heredity. Secondary Cutis Laxa occurs later in life [6]. In both the hereditary or acquired types, the internal organs are frequently involved [7]. In previous study a baby with Cutis Laxa who underwent herniorraphy with pulmonary stenosis under general anesthesia reported [8]. In this case report, the baby with Cutis Laxa had severe wheeze

and respiratory distress after extubation. Immediately the trachea was reintubated and drugs were given for bronchospasm [6]. We consider that these patients usually come to the pediatric, orthopedic or plastic surgeons for the treatment of this disorder or its associated problems [9]. A few cases of Cutis Laxa have been reported [10]. A baby out of five who underwent surgery (for congenital lobar emphysema and lung cyst under general anesthesia) has been reported. This baby had severe wheeze and intercostal in drawing after extubation. Immediately trachea was reintubated. After 48 hours, endotracheal tube was blocked and removed. The neonatologist who was consulted, noted other anomalies namely hypertelorism, short columella, hook nose, large philtrum, large upper lip with loose skin on dorsum of hands and back of the neck. The diagnosis was "Cutis Laxa" with systemic involvement. There are also case reports of association of Cutis Laxa with pulmonary emphysema [6].

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