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Case report

Successful anesthetic and airway management in Coffin-Siris syndrome with congenital heart disease: Case report

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KEYWORDS

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Abstract *Introduction:* Coffin-Siris Syndrome (CSS) is a rare congenital malformation syndrome characterized with mild to severe developmental and cognitive delay, coarse facial features, fifth digit aplasia or hypoplasia associated with ectodermal, constitutional and organ-related (cardiac/neurological/gastrointestinal/genitourinary...) anomalies. Here, we have reported a successful anesthetic and airway management in a case of 5-year old boy with CSS who underwent congenital heart surgery.
Case report: A 5-year old male child weighing 14 kg, who was diagnosed as CSS underwent operation for the repair of partial atrioventricular septal defect and secundum atrial septal defect. This case report pertains to the successful anesthetic and airway management in the background of difficult airway and presence of various cardiac abnormalities.
Although patient was anticipated to be difficult for intubation due to laryngomalacia, micrognathia, macroglossia, tracheal intubation was performed without any difficulty using fiber-optic laryngoscopy. At the end of the operation, the patient was transferred to the cardiovascular intensive care unit and was extubated when his spontaneous breathing was satisfactory 4 h later after the operation without any complication.
Results and discussion: CSS often requires surgery and anesthetic intervention. The abnormal facial and airway as well as mental related features may lead intubation difficult, potentially due to short neck, large tongue and lips, poor dentition and poor communication.
Thinking that the practicing anesthetist needs to have appropriate knowledge for this entity and the equipment for managing difficult airway should readily be available. One of these patients which successfully managed without any complication was described in this brief report.

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1. Introduction

Coffin-Siris Syndrome (CSS), known as "fifth digit syndrome" is a rare congenital anomaly characterized by many variable signs and symptoms, varying degree of intellectual and motor developmental problems, "coarse" facial features, abnormalities of the fifth fingers; cardiac/neurological/gastrointestinal/genitourinary anomalies. Respiratory infections and difficult feeding are common during infancy [1–3].

Congenital heart abnormalities are also heterogeneous, observed more than 30% of cases [4].

Airway management can be difficult because micrognathia, macroglossia, hypotonia, and lax joint can also be the problem as well as mental status due to poor communication [2].

This report presents a successful anesthetic and airway management in a 5-year old boy with CSS who underwent congenital heart surgery.

2. Case report

A five-years-old boy with mental retardation, a height of 109 cm and weighs 14 kg having CSS was admitted to our hospital for the operation of congenital heart disease. At birth he had hypothyroidism, nephrocalcinosis, respiratory problems including snoring, noisy breathing, change of voice, recurrent croup, sleep apnea, partial atrioventricular septal defect (AVSD) with secundum atrial septal defect (ASD). Recurrent respiratory infections due to aspiration persisted throughout early childhood and feeding difficulties were present. Mental development was also significantly retarded.

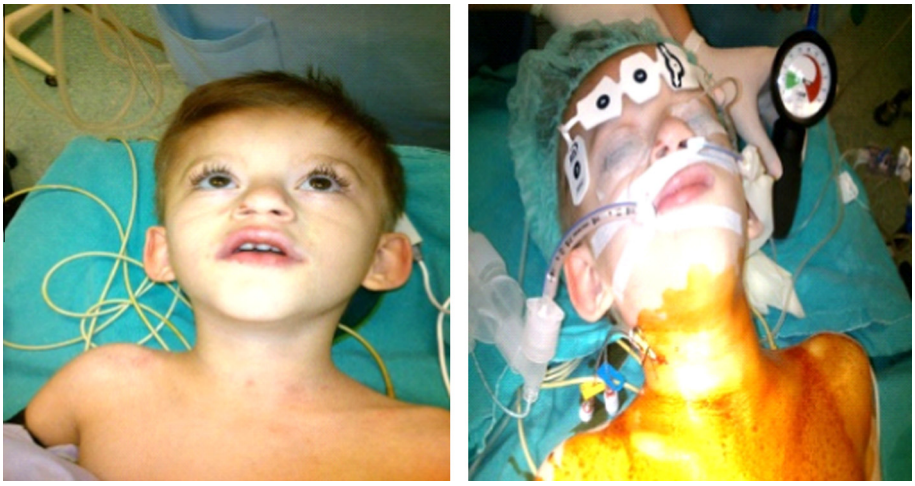
Physical examination revealed three major must-have features of CSS, namely retarded mental and cognitive growth; coarse facies reflected by flat nasal bridge, abnormal ears, wide mouth, microglossia, full lips and micrognathia; hypoplastic fifth finger terminal phalanx and the three minor findings of the syndrome (hypotonia, pectus carinatus and laryngomalacia). Muscles were hypotonic and pectus carinatus was prominent (Fig. 1). The patient was scheduled for cardiac surgery for his cardiac abnormalities, partial AVSD and secundum ASD.

After premedication with midazolam (0.05 mg/kg IV) the patient was admitted to operating room. Continuous monitoring was performed by evaluation of noninvasive blood pressure, pulse-oximetry and electrocardiography. His initial vital signs showed blood pressure (BP) of 91/57 mmHg, pulse rate of 113 beats/min, respiratory rate of 31/min and oxygen saturation rate of 99%, and skin temperature was 36 °C.

General anesthesia was initiated with 2 mcg/kg/min remifentanil infusion. Five minutes later, after establishing successful bag mask ventilation fiber-optic bronchoscopy (FOB) was carried out. Cuffed endotracheal tube number 5.5 mm internal diameter was safely placed into the trachea without trouble. Endotracheal cuff pressure was maintained among 10–15 cm H_2O which was continuously monitored till extubation (Fig. 2). After confirming effective endotracheal intubation, sodium thiopental 4 mg/kg and rocuronium 0.6 mg/kg were given. Anesthesia was maintained with isoflurane 50% oxygen/air and continuous infusion of remifentanil (0.01–1 mcg/kg/min) until the end of surgical procedures with the aim of keeping index of consciousness (IOC) values within 40–60, heart rate and blood pressure within the 30% range.

Depth of anesthesia was monitored with index of consciousness (IOC, Morpheus Medical, Spain) [K]. Cerebral (rSO-C) and somatic (rSOS) tissue oxygen saturation were monitored, were stable, and did not change compared to the initial values during operation (Fig. 3). Data were continuously updated at two readings per second and average recordings saved at 1 min intervals (Pediatric SomaSensor, Model SPFB, for children 4–40 kg by Somanetics Corporation, Troy, Michigan for the INVOS 5100 Cerebral oximeter). Operation and anesthesia were uneventful. Remifentanil was preferred for slow induction of anesthesia being advantageous for hemodynamic stability in this case.

After operation, he was transferred to cardiovascular intensive care unit (CICU) where remifentanil infusion and IOC monitoring were continued. Sedation level was assessed by Ramsay Sedation Score (RSS), and pain level was evaluated by Children's Hospital Eastern Ontario Pain Scale (CHEOPS). In CICU, the patient was connected to Servo 300 ventilator (Siemens-Elema, Solna, Sweden). He received time-cycled SIMV volume control mode. When he remained stable



Figures 1–2 Facial features of Coffin-Siris Syndrome.

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