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Airway Management in a Patient with Klippel-Feil Syndrome Using Extracorporeal Membrane Oxygenator

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Case Study

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ABSTRACT

We present a case of very difficult airway management resulting from a rare syndrome (Klippel-Feil) and rheumatic heart disease requiring a double valve replacement. The difficult airway consisted of a combination of difficult intubation, very difficult ventilation, and an almost impossible surgical airway. The only solution we found for such a complicated setup was the usage of Extracorporeal Membrane Oxygenator (ECMO) as a tool to allow the surgical construction of a safe airway. ECMO proved itself as a safe and efficient tool for securing the patient's hemodynamic and respiratory status. It should be remembered in such complicated clinical situations.

ABBREVIATIONS

ECMO: Extracorporeal Membrane Oxygenator; KFS: Klippel-Feil Syndrome; ENT: Ear-Nose-Throat

INTRODUCTION

Klippel-Feil syndrome (KFS) is a rare skeletal disorder affecting one of 42,000 births, with 55% female's predominance. The syndrome is primarily characterized by abnormal union or fusion of two or more bones of the spinal vertebrae. Some affected individuals may also have an abnormally short neck, restricted movement of the head and neck and a low hairline at the back of the head (posterior hairline). KFS patients requiring surgical intervention constitute an unusual high-risk group for anesthesia management. Due to the abnormally restricted motion of the cervical vertebra, airway management of these patients is extremely difficult. Not only that endotracheal intubation is very difficult in these patients, but also surgical airway access is almost impossible due to the lack of anterior access to the larynx or trachea [1].

Extracorporeal membrane oxygenator (ECMO) is an advanced form of life support technology whereby venous blood is oxygenated outside of the body and returned to the patient. ECMO was initially used as last-resort rescue therapy for patients with severe respiratory failure. Over the last four decades, it has developed into a safe, standard therapy for new born babies with progressive cardiorespiratory failure, as a resuscitation therapy after cardiac arrest, and in combination with other treatments such as hypothermia and various blood filtration therapies. ECMO has also become routine for children and adults with all forms of cardiogenic shock. As ECMO equipment becomes safer, earlier use improves patient outcomes. ECMO is very effective in maintaining adequate oxygenation and ventilation of the blood, while providing adequate cardiac output at the same time. It can be used either in venoarterial or in venovenous modes. ECMO can provide a short-term support for allowing the establishment of secure airway and proper ventilation and oxygenation [2-4].

CASE DESCRIPTION

We describe a case of a 20 years old male with type I KFS (characterized by complete fusion of the cervical vertebrae) and rheumatic heart disease [5]. The patient was admitted to the hospital due to rapid atrial flutter (150 beats/min with a 2:1 AV

conduction) and subsequent pulmonary edema. After initial stabilization and treatment of his atrial flutter and pulmonary edema, he underwent a complete workup in the cardiology department. The workup included echocardiography which demonstrated preserved right and left ventricular ejection fraction, a severely dilated left atrium (5.2 cm diameter, 35 cm^2 end-systolic area), a severely stenotic mitral valve with a mean pressure gradient of 22 mm Hg and a mitral valve area of 0.6 cm^2 **(Figure 1)**.

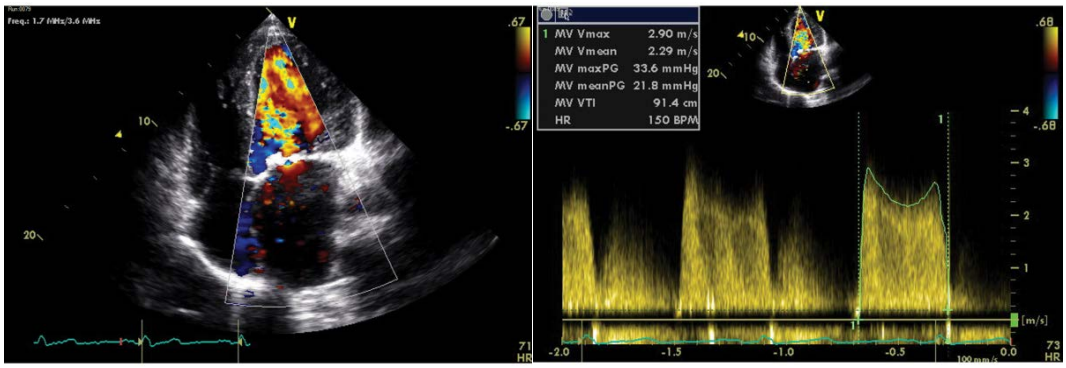

Figure 1. Mitral valve stenosis and mean gradient.

The aortic valve was severely regurgitant with mild-moderate stenosis (mean pressure gradient of 36mmHg and an indexed aortic valve area of 0.83 cm^2/m^2) **(Figure 2)**.

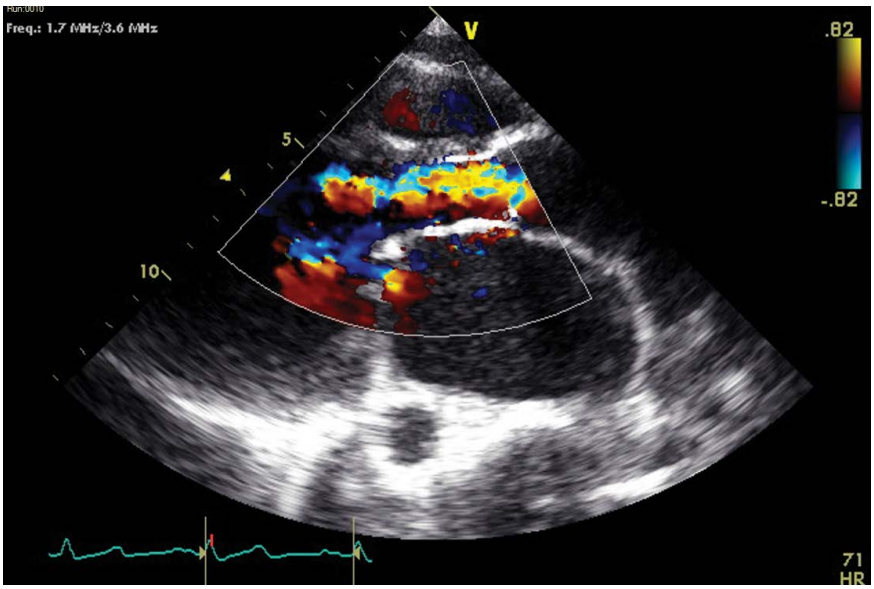

Figure 2. Severely regurgitant aortic valve.

Due to the clinical presentation and the echocardiographic findings, the patient was scheduled for an elective double valve replacement: mitral and aortic valves with mechanical valves. A preoperative anesthesiology consult was held. The patient's clinical syndrome led to a fixed position of his chin over his chest, with almost no anterio-posterior mobility (flexion-extension). Accordingly, all precautions for a difficult airway management were made, including fiberoptic laryngoscopy. On the operative day, due to the severe mitral stenosis and recent pulmonary edema, an awake intubation was not attempted, and a decision was made to attempt a regular intubation in a difficult airway patient. After induction of anesthesia, a first attempt at direct laryngoscopy and intubation was made, but failed. Subsequently, the patient developed a severe bronchospasm which led to difficult mask ventilation. An attempt at fiberoptic laryngoscopy was made but again failed, due to the complete lack of neck extension and laryngospasm. Finally, a laryngeal airway mask was inserted to allow oxygenation and ventilation. Concurrently, the patient developed severe pulmonary edema and the ventilation became extremely difficult. A team of Ear-Nose-Throat (ENT) surgeons was summoned urgently to attempt establishing a surgical airway. The only possible anatomical access to the larynx was through the lateral neck. However, due to the deteriorating respiratory status of the patient and the extremely difficult anatomical exposure, performing such a surgery was not feasible. During the entire time, the patient had relatively stable and preserved oxygenation but poor ventilation, with reserved PO_2 and a very high PCO_2. At that time, a decision was made to postpone the planned surgery and to zonnect the patient to an ECMO in order to allow the ENT surgeons to perform the surgically difficult irway. The groin of the patient was prepped and draped urgently and the right femoral vessels dissected and cannulated after heparinization. A 17F cannula was inserted into the femoral artery and a 21F venous cannula was inserted into the femoral vein to the right atrium [5-7]. A veno-arterial ECMO circuit was established and the patient was stabilized enough to allow cessation of

airway ventilation. This provided a clear and safe possibility for the ENT team to create a left-lateral neck tracheostomy through an open approach **(Figures 3 and 4)**.

Figure 3. Tracheostomy in the lateral neck.

Figure 4. Tracheostomy in the lateral neck.

The surgical procedure lasted 50 min and this allowed an effective and safe oxygenation and ventilation. It took another half hour to wean the patient from ECMO. The femoral vessels were repaired and groin incision closed. The patient was later transferred back to the cardiac intensive care unit. Due to the suspicion of prolonged hypercapnia and hypoxia which took place during the attempts to achieve a definite airway, we decided to cool the patient to 35°C for 48 h.

DISCUSSION

Two days later, he was warmed up again and allowed to wake up. Once his neurological status was declared to be intact, weaning from ventilation was performed. The patient was allowed to recover from the event and, 5 days later he was taken back to the operating room, for replacement of his aortic and mitral valves. A 21 mm aortic valve and a 29 mm mechanical mitral valve were implanted. The operation was uneventful and the patient was weaned from ventilation 24 h after surgery. After a successful postoperative course, he was discharged home on postoperative day 11. A CT scan was performed before discharge and demonstrated the severe fixation and union of cervical vertebrae **(Figures 5 and 6)**.

Figure 5. Reconstructed CT image demonstrating the union of the cervical vertebrae.

Figure 6. Reconstructed CT image demonstrating the union of the cervical vertebrae.

His pre-discharge echocardiography demonstrated normally functioning aortic and mitral prostheses.

CONCLUSION

We present a case of very difficult airway management resulting from a rare syndrome (Klippel-Feil) and rheumatic heart disease requiring a double valve replacement. The difficult airway consisted of a combination of difficult intubation, very difficult ventilation (due to mitral stenosis), and an almost impossible surgical airway. The only solution we found for such a complicated setup was the usage of ECMO as a tool to allow the surgical construction of a safe airway. ECMO proved itself as a safe and efficient tool for securing the patient's hemodynamic and respiratory status. ECMO is an important tool in the armamentarium of the cardiac surgeon and the anesthesiologist and intensivists. ECMO should be remembered in such complicated clinical situations of difficult airway and should be an internal part of the “difficult airway” emergency management protocol, especially when managing patients with cervical vertebral fusion such as Klippel-Feil syndrome.

CONSENT

The patient has given his consent for the case report to be published.

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