

## Dorsal Spinal Arthrodesis in a Patient with Tetra-Amelia: A Case Report

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### Abstract

**Background and purpose:** Tetra-amelia is a syndrome characterized by absence (complete) for all four members and anomalies. The objective of this report was to describe the case of a patient with tetra-amelia underwent arthrodesis of spine to correct severe scoliosis.

**Case report:** Male patient-12 years old, 41.1 kg, carrying tetra-amelia, underwent surgery to correct spine with severe scoliosis spine arthrodesis between T4 and L1.

**Discussion:** The management of the patient with tetra-amelia becomes a challenge for the implications of the airway, intraoperative monitoring, fluid replacement and response to vasoactive drugs. This syndrome is uncommon and rare reports in the literature also.

**Keywords:** Tetra-amelia; Case report; Anesthesiology; Monitoring; Intraoperative; Intraoperative Period

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### Introduction

Tetra-amelia is a syndrome characterized by absence (complete) for all four members and anomalies involving the skull and face (cleft lip/cleft palate, micrognathia, microtia, no nose septum, choanal atresia, absence of nose), eyes (microphthalmia, microcornea, cataracts, coloboma, eyelid fusion), urogenital system (renal agenesis, persistent cloaca, no external genitalia, atresia of the vagina), anus (atresia), heart, lungs (hypoplasia/aplasia), the skeleton (hypoplasia/absence of pelvic bones, no ribs, no vertebrae) and the system central nervous (agenesis of the olfactory nerves, agenesis of the optic nerves, agenesis of the corpus callosum, hydrocephalus). Affected infants are often stillborn or die shortly after birth [1,2].

The objective of this report was to describe the case of a patient with tetra-amelia submitted to arthrodesis of spine to correct severe scoliosis.

### Case Report

Male patient, 12 years and 8 months old, weight 41.1 kilograms, 91 cm in height, carrier tetra-amelia (absence of all four limbs) underwent surgery for correction of spine severe scoliosis with posterior spinal fusion from T4 to L1 (Figure 1).

During pre-anesthetic evaluation, presented in good condition, hydrated, no fever and eupneic. Normal psychomotor

development and no other comorbidities. Moved to their way. Denied allergies, addictions and family and personal history negative for complications anesthetic. Evaluation Mallampati class I with good mouth opening and class II according to American Society of Anesthesiologists (ASA). Normal cardiac and pulmonary auscultation. Initial tests showed hematocrit of 39.7%, hemoglobin of 13.7 g/dL, leukocytes of 8.350/mm<sup>3</sup>, 232.000/mm<sup>3</sup> platelets, serum sodium of 136.1 mEq/L, serum potassium of 4.2 mEq/L, fasting blood glucose of 89 mg/dL, creatinine of 0.67 mg/dL, urea of 13 mg/dL, activation time prothrombin of 100.3% and normal electrocardiogram.

Chest X-ray with significant deformity thoracic, cardiac and pulmonary parenchyma area no peculiarities. The patient was placed supine, monitored, and pre medicated with midazolam 5 mg intravenous (IV). After pre-oxygenation, anesthesia was induced with atropine 0.25 mg fentanyl 150 µg IV propofol 90 mg IV and 20 mg atracurium IV.

The laryngoscope blade was introduced into the oral cavity from the right, with leave of the language and try to visualize the

epiglottis, being characterized as Cormack-Lehane grade II and intubation was successful.

After intubation was punctured right subclavian vein for pressure maintenance central venous and arterial vascular surgeon performed dissection in the left upper limb stump to maintain mean arterial pressure.

The monitoring was maintained sinus rhythm, saturation between 99% and 100%, end tidal CO<sub>2</sub> between 28 and 33 and mean arterial pressure between 65 and 80 throughout the period. Diuresis total was 250 ml (0.8 ml.kg<sup>-1</sup>.h<sup>-1</sup>). The intraoperative blood gases and pH showed immediate postoperative between 7.32 and 7.37; PO<sub>2</sub> between 98 and 121, between 21.4 and 24.6 bicarbonate and base excess between -3.4 and +2.1.

There were two spare packed red blood cells (700 ml) and a bag of fresh plasma and the use of tranexamic acid infusion pump 40 mg.h<sup>-1</sup> for two hours and a single dose of clonidine 50 mcg IV.

Anesthesia was maintained with isoflurane and remifentanyl between 0.1 and 0.25 µg.kg<sup>-1</sup>.min and atracurium as an option for muscle relaxation. Total anesthesia time was 11 hours and 20 minutes and course proceeded without intraoperative hypotension or need for vasoactive drugs. The patient was extubated taken at surgery and sent to the Intensive Care Unit.

## Discussion

So far, this is the first case of tetra-amelia published in the literature submitted to general anesthesia, since previous studies have described cases with [3,4] to spinal block the anesthetic management.

The challenge in this type of case starts from the pre-anesthetic into the intra-and postoperative management, since the tetra-amelia is unusual and the description of the cases in the literature also rare. Most patients with this syndrome are stillborn or die

shortly after birth. The case described by Otsuka did not fit in this syndrome, because it was a patient with underlying cause of Buerger's disease [3].

The patient in this case was not investigated as to the cause of tetra-amelia. The concern with this patient's airway was anticipated, since palate and upper airway malformation is described in this syndrome [1,2]. However, access of airways was not uneventful.

Likewise are described malformation of the central nervous system, pulmonary, and urogenital, but in this case these malformations were not evident on clinical examination [1,2].

The psychomotor development was normal to auscultation and placement of the vesical catheter also successfully. It would be able to carry out a lung function evaluation by spirometry, however, scoliosis would also be a limiting factor in the evaluation of this function beyond the actual prediction of reference values for age according to the normal curve.

The blood volume loss calculation was done with weighing gauze and sponges, as well as measurement of the volumes aspirated, plus being more 5 ml.kg<sup>-1</sup>.h<sup>-1</sup> of insensible losses.

Another point of controversy was the difficulty in the way of calculating the blood volume as this patient has no extremities, leaving doubt whether the usual bases of calculation would be compatible, since it has a higher proportion of large caliber vessels than the rest of the population. Usually for calculation of blood volume using the ratio of 70 ml.kg<sup>-1</sup> model, although doubtful was applied in this case [5]. There was no need for vasoactive drugs, but this was another question about the behavior of these drugs to reduce the proportion of resistance vessels.

The management of the patient with tetra-amelia becomes a challenge for the implications of the airway, intraoperative monitoring, fluid replacement and response to vasoactive drugs. This syndrome is unusual and also rare reports in the literature.



**Figure 1** Male patient, underwent surgery for correction of spine severe scoliosis with posterior spinal fusion.

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