


# Anesthesia for preoperative non-invasive intracranial pressure measurement in a child with Apert syndrome: a case report

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**Introduction:** The presence of craniofacial alterations such as craniosynostosis and midface hypoplasia are some features of Apert syndrome. Those characteristics may lead to intracranial hypertension. For clinical evaluation, a non-invasive intracranial pressure measurement can be performed both as a preoperative examination and to evaluate surgical treatment. Several methods have been used to measure intracranial pressure non-invasively, such as optic nerve sheath ultrasound, optical coherence tomography, and intracranial pressure waveform (ICPw) monitoring with the sensor Brain4Care®. Some methods like ICPw, require that the patient remains standstill. Because the patient wasn't cooperative, it was decided to register the ICPw under hypnosis. A specific anesthetic planning was done to minimize the impact on the ICPw.

**Case presentation:** We present a 6 years old child with Apert Syndrome with ICPw registered by Brain4Care® device before and after monobloc craniofacial advancement with internal distraction. The chosen anesthetic was Sevoflurane at a dose  $\leq 0.9\%$  of inspired gases and air/oxygen proportion of 50/50%, with a laryngeal mask, under spontaneous ventilation, with normal end-tidal CO<sub>2</sub> range. The preoperative ICPw suggested low cranial compliance, with a mean P2/P1 index above 1.2. The postoperative showed a mean P2/P1 index under 1.2, demonstrating improved cranial compliance. The same anesthetic planning was done on both occasions.

**Conclusion:** Likely, an appropriate way to obtain a reliable non-invasive ICPw measurement under hypnosis includes positioning the patient at neutral decubitus, choosing hypnotics with minimal impact on ICP, and maintaining spontaneous ventilation during the exam.

**Keywords:** Acrocephalosyndactyly (Apert), Anesthesia, Intracranial pressure.

## INTRODUCTION

Apert syndrome is a congenital anomaly characterized by craniosynostosis, midface hypoplasia, and syndactyly (1). The first two characteristics may lead to intracranial hypertension (ICH) due to low cranial compliance, venous hypertension, and hypercapnia secondary to obstructive apnea (2). Yet, airway obstruction at various levels may cause difficult airway management during anesthesia (3). ICH in Apert patients can also be related to hydrocephalus (4). Elevated intracranial pressure (ICP) can cause cognitive and visual impairment, but ICP monitoring requires a surgical procedure that is not routinely indicated. Several methods have been used to measure ICP in a non-invasively way, such as optic nerve sheath ultrasound, optical coherence tomography, and ICP waveform (ICPw) recording with sensor Brain4Care® (B4C) (5). Although considered a non-invasive method, the B4C device sensor requires patient collaboration because it depends on monitoring skull oscillation, and patient movement may cause interference.

During the ICPw recording, the patient must stay still and minimize movement. This collaboration is not always possible, making the ICPw recording and the retinography difficult. Given the importance of assessing the ICP, the associated risk of invasive ICP monitoring, the availability of non-invasive technology, and the lack of patient collaboration, it was decided to register the ICPw under general anesthesia. It is known that anesthetics, intubation, and ventilation can affect ICP, so this report aims to demonstrate that it was possible to safely measure the ICPw under anesthesia in a patient with Apert syndrome.

## CASE REPORT

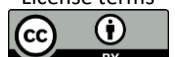
Male individual, seven years old, with Apert syndrome. He underwent posterior cranial decompression surgery at one year and nine months and had follow-ups with the craniofacial surgery team at the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC-USP). At age 6, a monoblock craniofacial advancement with an internal distractor was



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done due to snoring complaints and sleep apnea episodes. As part of the preoperative evaluation, the ICPw was registered by a B4C sensor the week before surgery, and retinography was registered using a portable Phelcom Eyer Maps® device. Due to the patient's lack of cooperation, the evaluations were conducted under general anesthesia. The patient was discharged the same day and underwent the surgical procedure the following week. On the day of the surgery, new second measurements were also done before the incision. The patient did not experience any complications during the period with the distractor, and five months later, the distractor was removed in the operating room. On this occasion, ICPw monitoring and retinography were performed for the third time. The initial curve suggested low cranial compliance, with a mean P2/P1 index of 1.38; after the monobloc advancement, the final measurement showed a mean P2/P1 index of 1.18 (Fig 1). An index P2/P1 below 1.2 is correlated with intracranial pressure below 20mmHg (6).

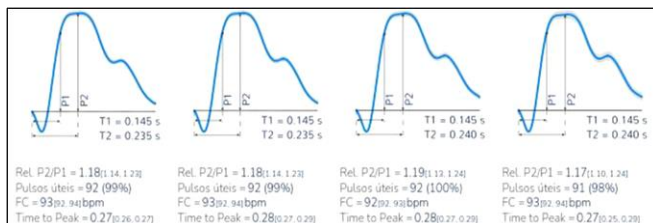


Figure 1- ICP waveform after monobloc craniofacial advancement

Retinography was performed by an ophthalmologist using a portable retinal camera Phelcom Eyer Maps®. Both exams pre and post-monobloc surgery showed no papillary edema or atrophy. (Fig 2).

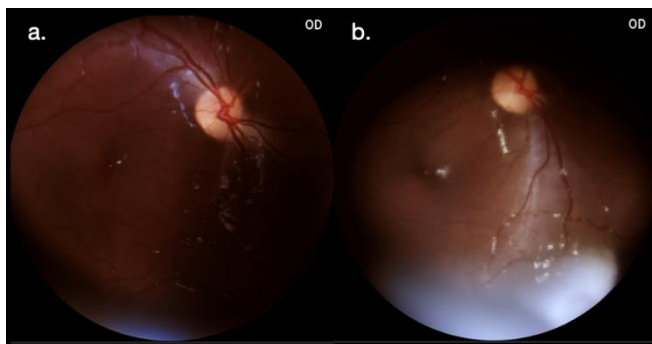


Figure 2- a. Retinography pre-monobloc advancement b. Retinography after advancement

The anesthesiologist was the same for all assessments and followed the same protocol to ensure anesthesia did not affect the curve measurement. The patient arrived at the surgical center fasting according to ASA protocols for age, alert, oriented, communicative, in good general condition, with no fever, without other changes to the physical examination, except for craniofacial malformation. No pre-anesthetic medication was administered before the

procedure. The individual was positioned in the horizontal dorsal decubitus and monitored with electrocardiography (ECG), pulse oximetry, and non-invasive blood pressure measurement. Then a face mask was administered with a mixture of 40% oxygen, 60% nitrous oxide, and 5% Sevoflurane. Shortly after, an appropriate level of consciousness was obtained, a peripheral intravenous line was installed, and a number 2.5 laryngeal mask was introduced, maintaining spontaneous ventilation without complications. Sevoflurane was then reduced to 0.9%, nitrous oxide was discontinued, and the diluent mixture of air and oxygen was held at 50%. The Dräger® Vamos® gas analyzer was used for gas concentration measurement. The patient remained positioned in the horizontal dorsal decubitus, and the B4C sensor was installed around the patient's head (figure 3).



Figure 3- Brain4Care® sensor device in place.

Peripheral oxygen saturation (SpO<sub>2</sub>), heart rate, non-invasive blood pressure, and gas analysis, including expired CO<sub>2</sub> (ETCO<sub>2</sub>), were monitored throughout the procedure (figure 4a). The airway was maintained with a laryngeal mask, and the patient was on spontaneous ventilation during the exam. Only after the Sevoflurane respiratory end-tidal level was less than 0,9% (figure 4b) did the B4C monitor start recording the ICPw variation curves for about 25 minutes. When B4C was online, there was no N<sub>2</sub>O detected at the gas analyzer. All hemodynamics and end-tidal CO<sub>2</sub> ranges were at normal values.

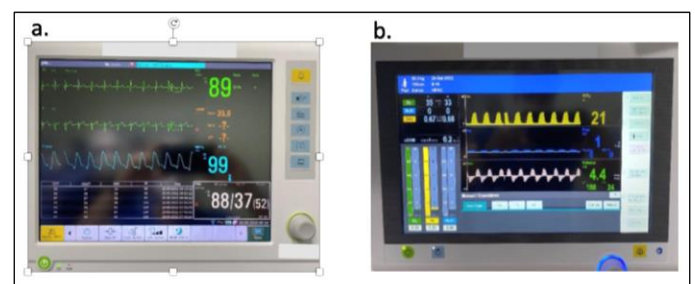


Figure 4- a. multiparameter monitoring; b. gas analysis.

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The inhalation anesthetic was discontinued after the measurement was concluded, and the laryngeal mask was removed for awakening. The following week, the same anesthesiologist did the same protocol, but at the end of the ICPw measurement, the laryngeal mask was replaced by a wired endotracheal tube, inhaled anesthesia was discontinued, and total intravenous anesthesia was initiated for the craniofacial surgery. At the end of the surgery, the individual was extubated in the recovery room and awakened with good post-anesthetic recovery. A third measurement was taken five months after surgery with the same protocol and anesthesiologist.

### DISCUSSION

The non-invasive measurement of ICPw using a B4C sensor has been used to evaluate ICP (6). This method may be beneficial for the evaluation of patients with craniosynostosis. But because of cognitive impairment and the patient's age, the lack of collaboration prevented ICPw adequate measurement with the B4C sensor. It was chosen to register the ICPw under hypnosis, even knowing that some specific anesthetic techniques can alter cerebral perfusion physiology and ICP (7). Drugs that may increase ICP were avoided, such as ketamine or nitrous oxide (8). Induction of hypnosis or maintenance with propofol and opioids can reduce cerebral metabolism and cerebral blood flow, thereby decreasing intracranial pressure(9). The minimum alveolar concentration (MAC) value is the concentration of an inhalation anesthetic agent in the lung alveoli required to prevent movement in response to a surgical stimulus in 50% of patients. It is known that inspired sevoflurane concentration at a greater than 1.0 MAC or 1,8% of inspired mixture gases may raise ICP by increasing cerebral blood flow (8). Therefore, Sevoflurane was chosen for induction and maintenance of hypnosis, with a dose during ICPw registration no greater than 0.5MAC or 0,9% to ensure no repercussions on ICP and no opioid was used. Because the inhalation anesthetic dose was very low, an endotracheal tube would not be tolerated and probably would elevate ICP by provoking airway reflexes. Considering these, a laryngeal mask was appropriate for the exam. In this study, pre-anesthetic midazolam was avoided as the individual was calm preoperatively, and there are reports of alterations in cerebral blood flow in specific situations (10).

Spontaneous ventilation is preferable since the use of controlled ventilation with or without positive end-expiratory pressure (PEEP) is known to reverse the physiological dynamics of respiratory flow, and there can be a decrease in cerebral venous return through the superior vena cava, increasing ICP (11). Another determining factor is the partial pressure of CO<sub>2</sub>. The patient was kept on spontaneous ventilation with normal levels of expired CO<sub>2</sub>. Hypercarbia can increase cerebral blood flow and ICP, whereas hyperventilation and hypocarbia could decrease ICP

(12). Likely, an appropriate way to obtain a reliable non-invasive ICPw measurement under hypnosis includes positioning the patient at neutral decubitus, choosing hypnotics with minimal impact on ICP, and maintaining spontaneous ventilation during the exam.

### CONCLUSION

We conclude that for a reliable NICEP measurement under hypnosis, the patient must be at neutral decubitus, choose hypnotics with minimal impact on ICP and maintain spontaneous ventilation during the exam.

### DISCLOSURES

#### *Ethical approval*

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the "Comite de Etica e Pesquisa HRAC CEP", CAAE:07704212.6.0000.0068 number: 4.537.300

#### *Consent to participate*

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

#### *Conflict of interest*

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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### CONTRIBUTIONS

- Luciano Brandao Machado: Conceptualization, Formal Analysis, Methodology, Writing – original draft
- Michele Madeira Brandao: Formal Analysis, Investigation, Writing – review & editing
- Andre Ferro: Data curation, Writing – original draft
- Tales Shinji Sawakuchi Minei: Data curation, Investigation
- Igor José Nogueira Gualberto: Data curation, Writing – original draft
- Nivaldo Alonso: Investigation, Supervision, Validation

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