SURGICAL MANAGEMENT OF PIERRE ROBIN SEQUENCE BY BEVERLI DOUGLAS´ TECHNIQUE. CASE REPORT.

MANEJO CIRÚRGICO DA SEQUÊNCIA DE PIERRE ROBIN PELA TÉCNICA DE BEVERLI DOUGLAS. RELATO DE CASO.

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ABSTRACT: A case of surgical management of Pierre Robin Sequence (PRS) by Beverli Douglas'technique is reported. The authors provide a short review of the literature and describe the choice procedure for respiratory airway relief in severe PRS as being safe and quick. The procedure performed (TLA) permits early discharge with a relatively good life condition, until definitive palatoplasty can be accomplished.

UNITERMS: Apnea, Cleft Palate, Glossoptosis, Micrognathia, Pierre Robin Sequence, Syndrome.

INTRODUCTION

In 1923, Pierre Robin, in a classic article "A Fall of the Base of the Tongue Considered as a New Cause of Nasopharyngeal Respiratory Impairment" introduced the term glossoptosis and emphasized the well-known respiratory complications of the condition that now bears his name. He stated that glossoptosis and the resulting respiratory obstruction was due to the "dysmorphic atresia of the mandible" (PALETTA *et al.*,1994).

Dr. Robin attributed failure to thrive and even death to respiratory and nutritional insufficiency, mainly caused by the tongue falling backward and

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downward in such a way as to obstruct upper airway passage. Palatal clefting was not mentioned in this initial description of the disease complex (FROHBERG & LANGE, 1993).

During the last few years, our knowledge of Pierre Robin syndrome has benefited from advances in embryonic neurobiology. It has been shown that the syndrome related to an anomalous development of the foetal brainstem. A multidisciplinary study makes it possible to value prospectively the various anomalies with which it may be associated and to decide on a coherent treatment. This type of management should reduce the morbidity and prevent the sequelae of Pierre Robin sequence (HUBERT *et al.*, 1991).

Robin sequence is now understood to be a grouping of clinical findings that does not represent a distinct multiple anomaly syndrome. Previously kown as the Pierre Robin syndrome, this fairly common association of micrognathia with cleft palate and upper airway obstruction was initially thought to be a specific disease, and entire treatment regimens were established to deal with presumed problems.Until recently, the management of Robin has not been excellent and mortality has been high. It is only a better understanding of the basic mechanisms leading to this sequence that has resulted in better care (SADEWITZ, 1992).

Infants with complications of PRS are at increased risk of airway obstruction and result hypoxia, cor pulmonale, failure to thrive, and cerebral impairment (BULL, GIVAN, SADOVE, 1990).

Though the problems associated with Robin sequence may be numerous, especially if the primary cause of the sequence is a multiple anomaly syndrome, the most acute problems in affected newborns is upper airway obstruction. Until recently it has been tacitly assumed that glossoptosis is always the cause of the airway obstruction. More recent evidence has shown that the sources of airway obstruction are multiple and the cause of apnea heterogeneous (SHER,1992).

Despite of all standard methods of achieving neonatal respiratory relief, such as nasopharingeal tubes (McEVITT, 1973; HEAF, HELMS, DINWIDDIE, 1982)., or lateral and prone positioning of the newborn (LEWIS & PASHAYAN, 1980), sometimes free respiratory flow cannot be achieved, being ventilatory support necessary. Oral intubation in these children is not an easy task, and most of the times they do not permit extubation in short time periods. The tracheostomy (MOYSON, 1961; JERESATY, HUSZAR, BASU, 1969; JOHNSON & TODD, 1980) has been the alternative of choice for those complicated patients in many services, but there are some considerations such as strictures, esophageal injury, cannula handling problems and so on.

The aim of the present paper is to remember an option for temporary respiratory relief, the Beverly Douglas´s technique (DOUGLAS, 1946), discussing its viability as a safe and liable procedure. This therapy allows postponement of a decision concerning corrective surgery.

CASE REPORT

A female newborn brought by cesarean section due to pelvic presentation and acute fetal distress, with a clinic gestational age of 35 weeks and 5 days, was transferred to the Hospital of Clinics of the Federal University of Uberlândia - MG with 3 hours of age, from another service due to respiratory distress.

Maternal history showed irregular prenatal follow-up, with no significant gestational problems. Mother´s age of 27, smoker, a box a day, weight gain of 10 Kg during gestation. Birth conditions were regular (Apgar score of 6 and 7 in the first and fifth minutes respectively), birth weight of 2550g, length of 46 cm. Premature amniorrhexis of 3 hours. In the first hour of life she presented with progressive respiratory distress, with generalized cyanosis.

At hospital admission the neonate presented with acute respiratory distress, with cyanosis, hypothermia, HR of 152 and BR of 72, presence of a posterior cleft palate, micrognathia and severe glossoptosis. No other gross findings. Initial measures for airway patency such as oral airway and lateral and prone positioning were attempted without success.

Due to the lack of a fiber neonatal bronchoscope, intubation showed extremely difficult, being emergencial traction of the tongue outwards with a 3-0 mononylon suture necessary in order to provide enough field to introduce the orotracheal tube. Right after intubation, the neonate became calm, with prompt relief of respiratory symptoms. Its immediate effectiveness was documented by repetitive blood gas analyses. She was kept in MV for 18 days, and Beverli Douglas glossopexy was indicated and performed.

On the fourth postoperative day the infant was extubated, and in the following day she was free of additional oxygen support. One month after the procedure, the child was in good conditions except for a mild suction impairment. An elective Stamm gastrotomy was performed and the infant was discharged at 55 days of age in good conditions, with cleft palate correction programmed.

GENETIC EVALUATION

The infant showed typical signs of Pierre Robin sequence, particularly micrognathia

(SHPRINTZEN, 1992), glossoptosis (PALETTA *et al.*, 1994) and a typical wide U-shaped cleft palate (GORLIN, CERVENKA, PRUZANSKY, 1971). Anomalous implantation of the 5th finger, thumbs aduction and palpable bilateral kidneys were observed.

It was suggested to exclude the 18 trissomy with cariotype examination and follow-up. The Stickler´s syndrome was also investigated by measurement of ocular problems in the family, ophtalmologic examination of the infant, upper members and backside x-rays, including wrist and hands.

Under the maxillofacial viewpoint, we proposed the glossopexy by Beverli Douglas´s technique. Before surgery, the infant presented other apnea and cyanosis episodes reversed by aspiration.

BEVERLI DOUGLAS´ TECHNIQUE

It was performed inhalatory general anesthesia with nasotracheal intubation using a 3 Portex tube without cuff. The local anesthesia with a small vasoconstrictor volume decreases the bleeding and facilitates the surgical management. The incision is made removing a strip of mucosa from inner lip and lower tongue portion.

Four landmarks, two in the tongue and two in the lower lip help holding the position duringthe surgery. The tongue is released from mandibular symphisis and tractioned outwards. The tongue mucosa suture attached to lip mucosa is made alternatively, from inner to outer, with 3-0 mononylon. A more resistant suture (0 mononylon) is used to perform a horizontal U, transfixing tongue and lower lip, being anchored by rubber tubes (ELY, 1980). On the fourth postoperative day the infant was extubated without intercorrences.

DISCUSSION

Although many respected authors advocate tracheostomy as the choice procedure for respiratory airway relief in severe PRS, one must be concerned that this is not a simple procedure, and unless it is performed by a skilled specialist, it may carry many complications due to the procedure itself, such as injuries to other neck structures as vessels, esophagus and thyroid, or late ones like strictures, many of them of difficult management. Another problem with tracheostomies in our experience is the lack of appropriate canals in developing countries like ours, what leads to improvisation and enhances the complications possibilities. TLA (tongue to lip adhesion), as described by Beverli Douglas, is a safe and quick procedure that permits early discharge with a relatively good life condition, until definitive palatoplasty can be accomplished.

Improvements in neonatal intensive postoperative intervention and home monitoring, the morbidity and mortality for children with PRS can be reduced markedly.

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RESUMO: Um caso de manejo cirúrgico da Sequência de Pierre Robin pela técnica de Beverli Douglas é relatado. Os autores fornecem uma breve revisão da literatura e descrevem o procedimento de escolha para o alívio das vias aéreas respiratórias na Sequência de Pierre Robin severa, como sendo seguro e rápido. O procedimento preconizado (TLA) permite uma alta hospitalar precoce, com uma condição de vida relativamente boa, até que uma palatoplastia definitiva seja realizada.

UNITERMOS: Apnéia, Glossoptose, Micrognatia, Palato Fissurado, Seqüência de Pierre Robin, Síndrome.

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Figure 1: Compensatory mechanism of glossoptosis by TLA (Beverli Douglas´ operation). A. The tongue retroposition blocks the upper airway similar to a ball´s valve. An U suture is performed transfixing the lip and the tongue aiming to lead the tongue outwards. B. The suture is attached on two rubber tubes, fixing the tongue to the lip (glossopexy). This maneuver corrects the respiratory obstruction due to glossoptosis. C. Mucosa peeling scheme. The four sutures of traction increase the visualisation of the field. D. After the fixation.