

Anaesthesia for a patient with Leigh's syndrome undergoing surgery for scoliosis

T. F. JACOBS (*), F. S. PLASSCHAERT (**), G. P. BOSSUYT (***), L. L. SZEGEDI (***) and L. L. HERREGODS (****)

Summary : Anaesthesia for patients with Leigh's syndrome has rarely been reported.

Leigh's syndrome or subacute necrotizing encephalomyelopathy is a neurodegenerative disorder of infancy or childhood. Acute exacerbation with respiratory failure may accompany surgery and general anaesthesia.

In this case report we describe the anaesthetic management of a 17 year old patient scheduled for spine surgery.

Key words : Anaesthesia ; Leigh's syndrome.

INTRODUCTION

Leigh's syndrome or subacute necrotizing encephalomyelopathy, described by Denis Leigh in 1951 (1), is a neurodegenerative disorder of infancy or childhood. The possible symptoms found within this syndrome are developmental delay, raised lactate, hypotonia, failure to thrive, poor feeding, ataxia, ophthalmoplegia, optic atrophy, seizures, respiratory disturbance and peripheral neuropathy (2).

Defects of pyruvate dehydrogenase complex, pyruvate carboxylase, cytochrome C oxidase, nicotinamide adenine dinucleotide Q reductase (NADH-Q reductase) have been associated with Leigh's syndrome (2, 3, 4).

Anaesthesia for patients with this syndrome has rarely been reported (5, 6). Acute exacerbation with respiratory failure may accompany surgery and general anaesthesia (7).

We hereby describe the anaesthetic course of a 17 year old patient, diagnosed with Leigh's syndrome, undergoing spine surgery.

CASE REPORT

A 17 year old female patient was scheduled for elective scoliosis surgery.

The patient (weight 19 kilograms, height 135 centimeters) had a proven cytochrome C oxidase deficiency and intermittent elevated serum lactate levels (32,8-42 mg/dl).

Reviewing her medical record revealed no peripartal problems. She was described as a hypotonic floppy child with failure to thrive and associated mental retardation.

Muscle biopsy showed some structural aberrations matching a mitochondrial metabolic disorder, and a too weak cytochrome C oxidase reaction.

At the age of two she was admitted to the hospital for aspiration pneumonia.

Brainstem evoked response audiometry (BERA) investigations at the age of four revealed severe hearing loss. She also had an important visual deficit.

Between the age of four and eight she was hospitalised seven times for upper respiratory tract infections and the associated metabolic acidosis.

A year ago she received a gastrostomy tube for feeding difficulties (swallowing problems and risk of aspiration).

CT-scan of the brain showed a loss of white matter and some degree of cerebellar atrophy.

This girl, suffering from a severe progressive neuromuscular scoliosis (The X-ray exam showed a C-shaped thoracolumbar scoliotic curve of 78 degrees measured between T8 and L5 ; there was a pelvic obliquity present of 14 degrees), was now scheduled for an operation that will correct her scoliosis (a posterior instrumentation (Isola – Depuy Acromed®) and spondylodesis from T2 to the pelvis using the Galvestone technique).

T. F. JACOBS ; F. S. PLASSCHAERT ; G. P. BOSSUYT ; L. L. SZEGEDI ; L. L. HERREGODS.

(*) Resident in anaesthesiology, Department of Anaesthesiology, Gent University Hospital, Gent, Belgium.

(**) Staff orthopaedic surgeon, Department of Orthopaedic Surgery Gent University Hospital, Gent, Belgium.

(***) Staff anaesthesiologist, (****) Professor in Anaesthesiology, Department of Anaesthesiology, Gent University Hospital, Gent, Belgium.

Address for correspondence : Dr. T. Jacobs, Gent University Hospital, Department of Anaesthesiology, De Pintelaan, 185, 9000 Gent, Belgium. E-mail : tom.jacobs@UGent.be.

The week before surgery she underwent cardiologic and respiratory investigations that were reassuring. Echocardiography showed a dilation of the aortic root (with suspicion of Marfan's syndrome), but revealed no other abnormalities. A nocturnal monitoring of saturation (SpO₂) turned out to be normal (time between 95-100% = 95%, 90-95% = 5%, %time < 90% = 0%, SpO₂ min = 91%, SpO₂ max = 100% and SpO₂ mean = 99%).

In order to minimise the risk of aspiration at the day of surgery she had been fasting overnight.

Induction of general anaesthesia was done with an inhalational mixture of sevoflurane (8%) in O₂/N₂O (70%/30%). In the meantime an 18G peripheral venous catheter was inserted at the right elbow. When this was in place we switched from inhalational anaesthesia to total intravenous anaesthesia (TIVA). Propofol (150-200 mcg/kg/min) and remifentanyl (0,10-0,20 mcg/kg/min) were administered continuously.

A single bolus of cisatracurium (6 mg) was given to facilitate intubation. A cuffed endotracheal tube n° 6 (Mallinckrodt Medical, Athlone, Ireland) was inserted and mechanical ventilation was started. We used the Dräger Julian (Dräger, Lübeck, Germany) mechanical ventilator to perform a volume controlled mechanical ventilation at constant flow, and a mixture of O₂/Air (FiO₂ : 0.4) was chosen. Inspiratory time was 33% of total time with an end inspiratory pause of 10% and a respiratory rate of 23. The ventilatory settings were kept constant throughout the intervention.

A nasogastric tube and an oesophageal temperature probe were also put into place. A central venous line (BD Secalon® Seldy 16G 20 cm single lumen) was placed in the right internal jugular vein. A 20G catheter was inserted in the right femoral artery, for continuous blood pressure monitoring.

Other intraoperative monitoring consisted of non-invasive arterial pressure (on the right arm), a three leads ECG, pulse-oximetry, capnography, and an oesophageal temperature probe. Diuresis could not be followed because of an early loss of a probably malpositioned urethral catheter. A red blood cell saving device (Cellsaver Electa, Dideco, Italy) was also installed. Before incision one gram of cefazoline was given intravenously.

The surgery was in prone position with cushions placed under the chest and pelvis of the patient. The procedure lasted about five hours. Estimated blood loss was approximately 550 ml.

A peroperative arterial blood gas analysis showed a pH of 7.42, a carbon dioxide partial pres-

sure (pCO₂) of 38 mmHg, an oxygen partial pressure (pO₂) of 201 mmHg, an oxygen saturation (SatO₂) of 100%, Na⁺ of 136 mmol/L, K⁺ of 2.9 mmol/L, Ca⁺⁺ of 0.85 mmol/L, lactate of 9 mg/dl, HCO₃⁻ of 24.6 mmol/L, Hct of 21%, Hb of 65 g/L. During surgery she received 2 liters of crystalloids (Plasmalyte A) and 500 ml of a colloid tetra starch solution. The washed content of the cell saving apparatus was then returned to the patient (114 ml of washed cells) and one unit of packed red cells was also given (323 ml).

For pain relief 600 milligrams of propacetamol and 50 milligrams of tramadol were administered. She also received two milligrams of morphine in bolus, one hour before the end of the surgery.

At the end of the procedure she was turned in the supine position where she spontaneously began breathing ten minutes after skin closure, and we successfully managed to extubate the trachea in the operating room (45 minutes after skin closure).

Owing to the severity of the surgical intervention and the fact that the patient had Leigh's syndrome, we decided to transfer her to the paediatric intensive care unit (PICU) for postoperative observation. During her PICU stay she received supplemental potassium (K⁺ was 3.8 mmol/L at discharge), and two units of fresh frozen plasma in order to correct a low PT. She also received supplemental oxygen via nasal prongs at a rate of 1 L/min, but this could be easily weaned the following morning. Haemodynamic and respiratory parameters and arterial blood gasses remained stable. No deterioration of neurological function occurred. The postoperative X-rays showed a vertebral column curve reduced to 22 degrees.

She was discharged the day after and transferred to the paediatric department of the hospital.

DISCUSSION

The diagnosis of Leigh's syndrome in this patient was mainly based on the combination of a proven cytochrome C oxidase deficiency, mental retardation, physical delay and other neurological manifestations (including hearing loss, visual deficits).

Although malignant hyperthermia is a rare syndrome, characterised by an acute hypermetabolic state within muscle tissue, we should always keep this in mind when we perform general anaesthesia in patients with a known musculoskeletal disease.

Since succinylcholine and volatile anaesthetics are considered to be the principal triggering

agents, they should be avoided in such cases. However, we decided to use sevoflurane for induction and encountered no problems.

We paid careful attention not to raise serum lactate levels. No exogenous lactate was administered, hence the choice of Plasmalyte A as crystalloid.

Our ventilator settings remained the same throughout surgery and resulted in a constant normocapnia.

Acute exacerbation with respiratory failure may follow general anaesthesia in patients with Leigh's syndrome. We used propacetamol and tramadol as well as morphine, which is known to depress spontaneous ventilation, for pain relief.

Emergence was without problems and the trachea could easily be extubated in the operating room. No respiratory failure occurred. Stay at the PICU was uneventful.

Since anaesthesiologic experiences with Leigh's syndrome are limited and recommendations are mainly based on case reports, careful attention should be paid to the type of general ana-

esthesia chosen. Meticulous review of the medical history as well as the preoperative investigations should help to select the proper anaesthetic agents.

References

1. Leigh D., *Subacute necrotizing encephalomyelopathy in an infant*, J. NEUROL. NEUROSURG. PSYCHIATRY, **14**, 216-21, 1951.
2. Rahman S., Blok R. B., Dahl H. H., *et al.*, *Leigh Syndrome : clinical features and biochemical and DNA abnormalities*, ANN. NEUROL., **39** (3), 343-51, 1996.
3. Kretzschmar H. A., DeArmond S. J., Koch T. K., *et al.*, *Pyruvate dehydrogenase complex deficiency as a cause of subacute necrotizing encephalopathy (Leigh disease)*, PEDIATRICS, **79**, 370-3, 1987.
4. DiMauro S., Servidei S., Zeviani M., *et al.*, *Cytochrome C oxidase deficiency in Leigh syndrome*, ANN. NEUROL., **22**, 498-506, 1987.
5. Ward D. S., *Anesthesia for a child with Leigh's syndrome*, ANESTHESIOLOGY, **55**, 80-1, 1981.
6. Shenkman Z., Krichevski I., Elpeleg O. N., *et al.*, *Anaesthetic management of a patient with Leigh's syndrome*, CAN. J. ANAESTH., **44** (10), 1091-5, 1997.
7. Grattan-Smith P. J., Shield L. K., Hopkins I. J., *et al.*, *Acute respiratory failure precipitated by general anesthesia in Leigh's syndrome*, J. CHILD NEUROL., **5**, 137-41, 1990.