

## Anaesthetic approach to a patient with Rett syndrome during tooth extraction

Ö Önal<sup>1\*</sup>, A Kaplan<sup>2</sup>

### Abstract

#### Introduction

Rett syndrome is a progressive developmental syndrome that occurs at the rate of 10,000–23,000 throughout the world and courses with autism, dementia, ataxia and loss of manual dexterity in girls. While children with Rett syndrome show a normal or near-normal development until the 6th to 18th months of their lives, they forget the skills they obtained with a rapid regression and then enter a long period of non-development. In this case report, a 26-year-old female patient undergoing operation for tooth extraction is presented.

#### Case report

26-year-old patient diagnosed with Rett syndrome was planned to undergo dental procedures under general anaesthesia. She had mental and motor developmental retardation and was not able to walk. Atrophy was present in all extremities with limitation in manual skills, bruxism and shouting episodes. For anaesthesia induction, propofol 3 mg kg<sup>-1</sup>, lidocain 1 mg kg<sup>-1</sup>, remifentanil 1 µg kg<sup>-1</sup> were used at loading dose and tracheal intubation was performed with muscular relaxation by rocuronium 0.6 mg kg<sup>-1</sup>. In maintenance, propofol and remifentanil infusion was made. Overall 14 teeth underwent procedure without any complications. At the end of the procedure,

propofol and remifentanil infusions were discontinued and patient extubated.

#### Conclusion

In patients with Rett syndrome, haemodynamic stabilisation should be maintained by vigilant monitoring in the perioperative period due to the presence of malignant hyperthermia risk, apnoea attacks that may occur during respiration and sleep, difficulty in intubation and extubation, prolongation of QT in ECG and abnormal T waves, which is important for safe anaesthesia.

#### Introduction

Rett syndrome (RS) is a progressive developmental syndrome that occurs at the rate of 10,000–23,000 throughout the world and courses with autism, dementia, ataxia and loss of manual dexterity in girls<sup>1</sup>. It is the second leading cause of mental retardation among girls following Down syndrome<sup>2</sup>. While children with RS show a normal or near-normal development until the 6th 18th months of their lives, they forget the skills they obtained with a rapid regression and then enter a long period of non-development. RS, which is X chromosome dominant, is a progressive neurodevelopmental disorder occurring in girls, was originally defined by Andreas Rett in 1966 as 'brain atrophy in childhood'<sup>3</sup>. Clinical picture includes respiratory disorders, EEG and ECG abnormalities, spasticity, advanced muscle breakdown and dystonia, peripheral motor dysfunction and scoliosis, which prompts caution in pre-anaesthesia preparation and administration of anaesthesia. In this case report, a 26-year-old female

patient undergoing operation for tooth extraction is presented.

#### Case report

A 26-year-old female patient diagnosed with RS was planned to undergo dental procedures under general anaesthesia owing to the many decayed teeth she has. She underwent preoperative evaluation. She was mentally retarded and her consciousness was open. However, she lacked cooperation and orientation. She had mental and motor developmental retardation and was not able to walk. Atrophy was present in all extremities, with limitation in manual, bruxism and shouting episodes. In cardiovascular examination, heart sounds were rhythmic without any additional sounds and murmur. In respiratory examination, there were no pathological findings except for minimal harshness in respiratory sounds. Laboratory tests were found to be normal. In ECG, prolongation in QT was found. Patients were evaluated by paediatric neurology, cardiology and chest diseases. Due to the risk of malignant hypothermia, dantrolene sodium was applied in accordance with the recommendations of paediatric neurology. The patient was taken to the operating theatre; saline infusion was initiated and ECG measurement, pulse oxymetry, axillary heart monitoring, non-invasive blood pressure and temperature probes were carried out. Vital parameters were heart rate 105/min, oxygen saturation on air 90% (100% on oxygen) and blood pressure 125/80 mmHg. For anaesthesia induction, propofol 3 mg/kg, lidocaine 1 mg/kg, remifentanil 1 µg/kg were used at loading dose, and tracheal intubation was performed with

\* Corresponding author  
Email: drozkanonal@selcuk.edu.tr

<sup>1</sup> Selçuk University Medical Faculty, Anesthesiology and Reanimation Department, Konya, Turkey

<sup>2</sup> Tepebaşı Teeth Hospital, Anesthesiology and Reanimation Department, Ankara, Turkey

muscular relaxation by rocuronium 0.6 mg/kg. In maintenance, propofol and remifentanyl infusion was made. Vital findings of the patient were stable during operation. Rectal temperature reached a maximum of 37.6°C during operation. Overall, 14 teeth underwent procedure without any complications. At the end of the procedure, propofol and remifentanyl infusions were discontinued, and the patient was extubated. For post-operative analgesia, the patient was administered 100 mg tramadol and 20 mg tenoxicam.

### Discussion

RS is a neurological condition originally described by Andreas Rett in 1966 which occurs mostly in girls<sup>4</sup>. It is characterised clinically by autistic behaviour, mental retardation, respiratory problems, loss of speech and manual skills, not being able to use hands purposefully, holding breath, oral-motor dysfunctions, gastrointestinal motility disorders, scoliosis, autonomous dysfunction and somatic developmental impairment<sup>5</sup>. Acquired elements of language disappeared and purposeful movement of the hands was lost, while stereotypic hand washing movements are shown<sup>6</sup>. Various movement disorders were exhibited, including dystonia, choreoathetosis, myoclonic jerk and stereotypic automatism<sup>6,7</sup>. Axial hypotonia and limb spasticity frequently develop. Cardiovascular, respiratory, neurological and metabolic features of this disorder have the potential of anaesthetic problems. Considerations resulting from neuromuscular involvement include lack of cooperation, control of seizures and limb movements, positioning and selection of muscle relaxants. Behavioural problems that can be associated with respiratory irregularities may be accelerated by medical manipulations or introduction to the operating room<sup>8</sup>. Deep preoperative sedation may prevent such a response but could have undesired side effects. Music is reported to have a soothing

effect in patients with RS and provide a temporary cessation of stereotypic movements<sup>9</sup>. The altered muscle tone demand and muscle wasting close attention to proper positioning. These children may be at risk of recurrent aspiration and impaired respiratory function due to muscle weakness and thoracic deformity. In addition, abnormal patterns of respiration are commonly seen<sup>10</sup>. Cirignotta et al. (8) have suggested that the frequent desaturations may cause permanent hypoxic damage and contribute to the progressive cerebral deterioration. The effect of anaesthetic agents on respiratory patterns during the perioperative period is yet to be known. The potential for aspiration, respiratory muscle weakness and apnoea should be considered in preoperative assessment of respiratory function. The patient presented had minimal respiratory symptoms, no evidence of recurrent aspiration and a normal chest x-ray. The severity of symptoms, nature of surgery and anaesthesia should be considered in the requirement for post-surgical admission to the ICU<sup>9</sup>. Vasomotor disturbances leading to cool limbs with tropic changes are observed in 50% of older affected children. Profuse sweating may also be seen<sup>9</sup>. Aggressive attempts are required in order to control heat loss.

In patients with RS, irregular respiration, tachypnea and apnoea attacks occur only when awake<sup>11</sup>. When it occurs together with rapidly progressive scoliosis, it may lead to severe respiratory failure in the patients. In neuromuscular scoliosis cases secondary to RS, muscular tonus loss occurs. It has been reported that in these patients serum potassium levels increase with the use of depolarising muscle relaxants such as succinylcholine<sup>12</sup>. In patients with scoliosis due to muscular dystrophy, susceptibility to malignant hyperthermia, cardiac dysrhythmia and side effects of succinylcholine increase. Hypermetabolic states observed in patients with RS indicate that anaesthesia

and anaesthetic agents may increase the risk of malignant hyperthermia. Cardiac arrhythmias associated with prolonged QT interval occur frequently in patients with RS. Drugs that can trigger cardiac arrhythmias and QT abnormalities should be avoided. In anaesthesia, the use of thiopental and succinylcholine, prokinetics such as sisaprid, antipsychotic such as tioridazin, tricyclic antidepressants such as imipramine, antiarrhythmics such as kinidin, amiodarone, sotalol as well as erythromycin and ketaconazole should be avoided<sup>13</sup>. As patients with RS are extremely sensitive to sedation drugs, postoperative recovery may be difficult. In addition, in patients with RS, due to the state of metabolic acidosis, the presence of low pain threshold, muscular degradation and the tendency of potassium level to be triggered by anaesthetic agents such as succinylcholine, the presence of abnormal EMG, loss of muscle, abnormal neural transmission and the risk of hyperthermia that can be triggered, prophylactic measures should be taken and operation should be carried out choosing the most suitable anaesthesia method.

### Conclusion

In patients with RS, haemodynamic stabilisation should be maintained by vigilant monitoring in the perioperative period due to the presence of malignant hyperthermia risk, apnoea attacks that may occur during respiration and sleep, difficulty in intubation and extubation, prolongation of QT in ECG and abnormal T waves, which is important for safe anaesthesia.

### Abbreviations

RS, Rett syndrome.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written

consent is available for review by the editor-in-chief of this journal.

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