Peculiar breathing in Rett syndrome: Anesthesiologist's nightmare

Dear Editor,

Rett syndrome is a rare, progressive neurological disorder due to degeneration of the central nervous system affecting only females,[1] having a prevalence of 1:25000.[2] Mental retardation, movement disorders, an abnormality in respiratory control, autistic behavior and seizures arise in childhood after a period of normal growth and development and pose a challenge to the anesthesiologist.[1,3]

A 4-year-old girl with Rett syndrome was scheduled for ophthalmic surgery for seborrheic dermatitis of the eyelid margin. Born by full term normal vaginal delivery, she had a delay in attainment of the developmental milestones. By the age of 2 years she lost the milestones, which she had attained. She had mental retardation, microcephaly, neuroregression with hypotonia, joint contraction [Figures 1 and 2], and squint. Her weight was 17 kg and blood pressure (BP) was 90/60 mmHg. Airway examination was unremarkable except for the caries in teeth [Figure 3]. Respiratory and cardiovascular systemic examinations were normal. Her hemoglobin was 10.4 g/dl and serum electrolytes were within normal limits. Electrocardiogram (ECG) showed T wave inversion in leads V_3-V_5 [Figure 4]. She was kept nil per oral adequately. In the operating room, her baseline vitals were heart rate of 118/min, BP of 88/60 mmHg and pulse oximetry showed 99% oxygen saturation. Since the procedure was expected to last about 20 min, our plan of anesthesia was general anesthesia with proseal Laryngeal mask airway (LMA) with assisted ventilation. After pre-oxygenation, 0.2 mg IV glycopyrrolate was given. Anesthesia was induced with titrating dose of 26 mg propofol, and 15 μg fentanyl. Mask ventilation was done with 2% sevoflurane in oxygen and size 2 proseal LMA was secured in place. She was kept on spontaneous ventilation with nitrous oxide, oxygen and sevoflurane. Intra operative period was uneventful, with no period of desaturation. At the end, inhaled anesthetics were tapered off, and the child was breathing well with 100% oxygen. Proseal LMA was removed once child was fully awake. After LMA removal, the respiration became irregular with diminished tidal volumes. There was a brief period of apnea which lasted for about 10 s, during which in spite of mask ventilation, the oxygen saturation dropped to 95%. The irregular breathing episodes were interspersed with regular breathing periods. This continued for about 20 min, during which we assisted the ventilation as and when required and saturation was maintained at 97-98%. After the child regained regular breathing with adequate tidal volumes, she was shifted to the high dependency unit and was kept under observation for next 8 h, while on oxygen via face mask at 4 L/min. Subsequent stay in the hospital was uneventful, and she was discharged the following day.

The features of Rett syndrome appear at the age of about 6-18 months, after a period of normal development.[2,4] The features of respiratory dysrhythmia, which includes hyperventilation and periodic apnea, gastrointestinal dysfunction, spasticity, joint contraction, diminished muscle tone, cardiac conduction abnormalities, T wave abnormalities and severe sinus bradycardia pose a challenge to the anesthesiologist.[5,6]

Pre-operative evaluation is crucial to analyze the breathing

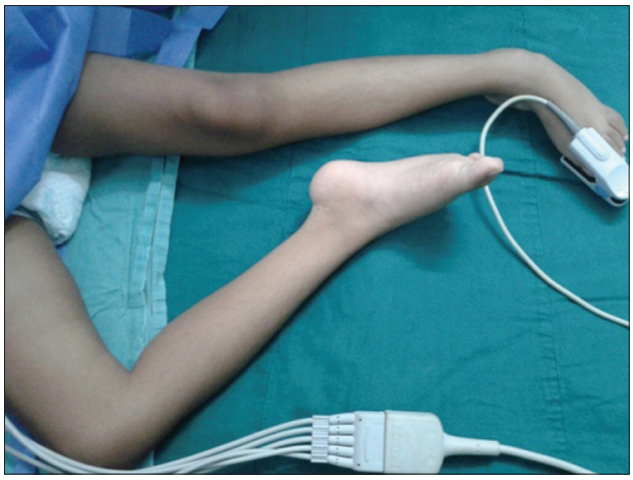

Figure 1: Lower limb contractures

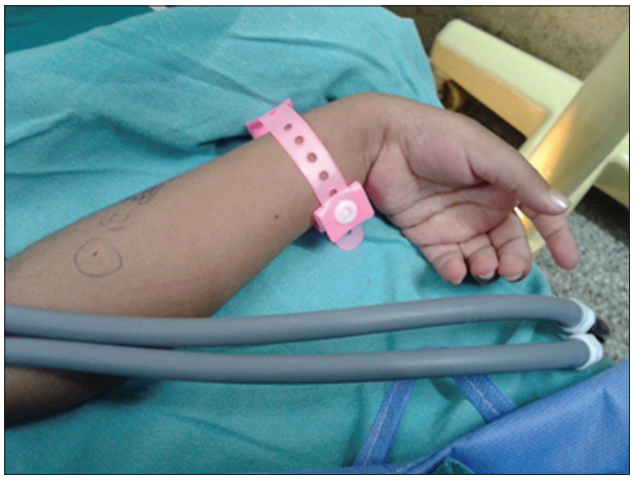

Figure 2: Contractures of hand

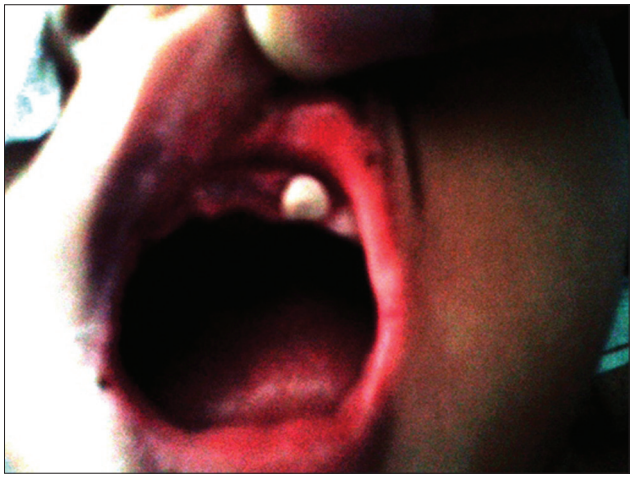

Figure 3: Caries and missing teeth in the child

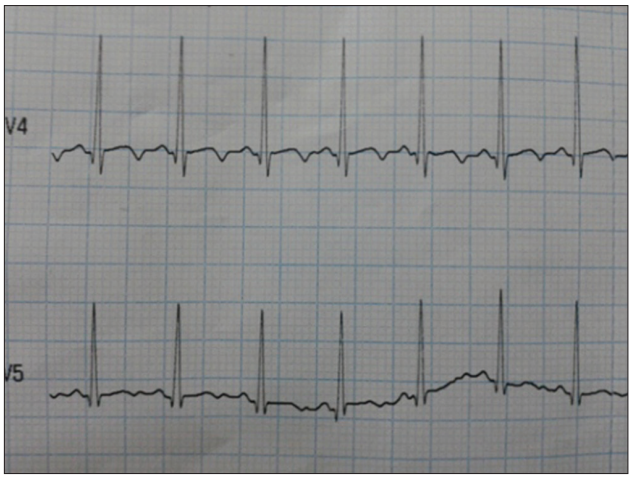

Figure 4: Electrocardiogram showing T wave inversion, a finding in Rett syndrome

patterns, arterial blood gases in case of any abnormality in breathing, presence of gastro intestinal reflux, autonomic disturbances, seizures, musculoskeletal disorders and analysis of ECG. Metabolic abnormalities in these patients include elevated ammonia concentration and lactic acidosis.[7] Intravenous fluid administration must be done with caution as acidosis may occur due to rapid IV fluid administration.[4]

These patients are excessively sensitive to sedative drugs, and delayed recovery from anesthesia has been reported.[8] Hence, we titrated the drugs to maintain optimal anesthetic depth and avoid overdose. Child was kept on spontaneous ventilation rather than controlled as that would require administration of more anesthetic agents.

Polygraphic studies show that episodes of irregular breathing and even apnea can occur during wakefulness, whereas no such irregularities occur during sleep, where breathing is stable. Wakefulness is associated with periods of disorganized, ineffective respiratory efforts, mixed with apnea.[9] The cause of this has been attributed to impairment of the behavioral control system of breathing, which is considered to be a forebrain function.[9,10]

We found this peculiar breathing pattern in our patient when she became fully awake and proseal LMA was removed. Though she had regularity in breathing while she was on spontaneous ventilation under anesthesia, it became irregular along with a brief period of apnea once she was awake.

Arterial desaturation and loss of consciousness during episodes of irregular breathing can also lead to permanent hypoxic damage.[9]

Though, there are reports of delayed recovery from anesthesia in cases of Rett syndrome, there is a paucity of reports describing this irregularity in breathing in a child who was otherwise fully awake and recovered from anesthesia. Even if a history of irregular breathing and apnea is not elicited in a patient, we should always anticipate respiratory impairment in these patients.[4] Intense monitoring in post-operative period is essential as frequent desaturations in these patients may cause progressive cerebral damage.[9]

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