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Respiratory Flutter Syndrome in a Neonate

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Respiratory flutter or diaphragmatic flutter is a rare disorder characterized by involuntary, high frequency contractions of the diaphragm. Only 7 cases are reported in infants till date. The present case presented with life threatening respiratory distress immediately after birth. In view of high respiratory rate of 120-154 per minute, clinical and fluoroscopic evidence of diaphragmatic contraction and absence of any obvious CNS, cardiovascular and respiratory pathology, respiratory flutter was diagnosed. It was also associated with dysphagia and laryngomalacia. The patient was managed with prolonged continuous positive airway pressures (CPAP) with partial success, but symptoms improved with use of chlorpromazine.

Key words: *Diaphragmatic flutter, Newborn, Respiratory distress.*

Respiratory or diaphragmatic flutter (DF) is a heterogeneous neurologic disorder, rarely reported in neonates, infants and children. It is characterized by rapid, involuntary high frequency contractions of the diaphragm (35-480/min), often superimposed on normal diaphragmatic excursion; and considered to be a form of myoclonus [1-3]. The reported respiratory patterns of DF are highly variable. It may occur in inspiration, expiration or in both phases of respiration, generally asynchronous with heart rate and usually resolves during sleep, but may persist within all stages of sleep. The duration of DF may vary from days to 18 years but typical to this disease, gas exchange (CO_2 and O_2) abnormalities are absent. Since the first case report in 1723, a case of self diagnosis by Leeuwenhoek [4], only 7 cases are reported in infants [2, 3, 5]. We report an infant with diaphragmatic flutter associated with dysphagia and laryngomalacia.

CASE REPORT

This full term male baby was delivered by elective caesarian section with birthweight of 3100g to a non-consanguineously married 23 years old primipara with no antenatal risk factors and normal antenatal ultrasound scans. Baby cried immediately after birth. At 2 hours of life, baby was shifted to NICU for excessive drooling of saliva and respiratory distress. Baby was intubated and ventilated at the referral NICU for 5 days, but in view of extubation failure twice, he was referred to our unit on 6th day of life. On examination, baby was hemodynamically stable and systemic examination was normal. He was lethargic, tone, and reflexes were poor, and pupils were normal. Baby was ventilated on SIMV mode, weaned and extubated to CPAP over next 48 hours. During ventilation, his respiratory rate was very high, irrespective of SIMV rate and level of sedation. In view of history of extubation failure, retrognathia and excessive pooling of secretions and recurrent upper lobe collapse of lungs, airway evaluation was done. Direct laryngoscopy revealed moderate laryngomalacia and omega shaped epiglottis; bronchoscopy was normal. Expanded newborn screening with immunoreactive trypsinogen (IRT) was normal. MRI brain, nerve conduction study and electromyography were normal. Sepsis work-up was negative. 2D echo cardiography was normal. During CPAP, respiratory distress persisted with a biphasic pattern of respiratory rate ranging from 40 – 154 per minute. Excessive secretions and respiratory distress was persistent even after 10 days of CPAP use. During clinical observation, it was noted that baby was "appearing frightened". Respiratory rate monitored with impendence plethysmography (Philips Intellivue MP 40

monitor) revealed a respiratory rate of >150 per minute. Video recording of respiratory movement showed high frequency like wiggle of chest and abdomen with interposed normal breathing. Video-fluoroscopic study showed a breathing rate of 160 breaths per minute with oscillatory movement of both sides of diaphragm. It also revealed oropharyngeal dysphagia characterized by a delay in swallow onset and silent aspiration in absence of cough and slow progression of dye in esophagus. Secretions decreased with glycopyrrolate but respiratory distress persisted. Characteristically, through out the course of 10 days of CPAP, arterial blood gases (ABG) were normal.

In view of high frequency intermittent contraction of diaphragm as evidenced by clinical observation impendence plethysmography monitor and fluoroscopy recording, diaphragmatic flutter was diagnosed. In view of poor response to continuous CPAP use, chlorpromazine in a dose of 1.5mg/kg/day in three divided doses was started through nasogastric tube. Clinical improvement was noticed over next 48 hours and infant was weaned from CPAP. Follow-up at 1 month was normal with no recurrence. Subsequently the child was lost to follow-up.

DISCUSSION

Diagnosis of DF is by visual observation of diaphragmatic flutter, fluoroscopic imaging, impendence pneumography, respiratory inductive plethysmography (RIP) and diaphragmatic electromyography. DF is difficult to diagnose in infants by physical examination owing to the normal rapid respiratory rate of infants and difficulty in differentiating superimposed flutter waves which may be in the frequency of heart rate. Fluoroscopy can differentiate diaphragmatic contrations from heart rate [5]. RIP with a frequency response up to 1800 per minute accurately diagnoses high frequency DF. Impendence pneumography, though commonly used in infants, as in our case, has limitation that it may not detect respiratory rate above 180 per minute.

Three types of breathing pattern have been reported in DF. These are tachypnea, dirythmic breathing superimposed with high frequency waves and with apnea [6]. In the present case high frequency intermittent contraction of the diaphragm superimposed on ordinary respiratory rhythm *i.e.* "dual rhythms", confirmed by fluoroscopy and being asynchronous with heart rate ensured a diagnosis of diaphragmatic flutter.

A complex interaction between swallowing and medullary breathing centre in brainstem normally co-ordinates breathing and swallowing. Our infant had dysphagia supporting the concept of common brainstem dysfunction seen in these babies. However, normal MRI brain suggests functional rather than gross anatomic lesion. The response of DF to different therapies is variable. A range of therapies are described including diazepam, carbamazepine, phenytoin, chlorpromazine and even phrenic nerve-crush [2]. Poor response to CPAP prompted us to treat with chlorpromazine, a centrally acting dopamine antagonist. This treatment resulted in marked reduction in DF and allowed us to wean from CPAP within few hours. No side-effects of chlorpromazine were observed during follow-up.

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