

Rett's Syndrome Following Bronchopneumonia

Rett syndrome (RS), a neurodevelopment disorder is almost exclusively seen in girls. Diagnosis is essentially clinical and is based on characteristic clinical features. Though a genetically inherited disorder, environmental factors like fever can trigger the onset of this syndrome in genetically predisposed subjects(1). We present a 4 year old female child who was developmentally normal till 17 months of age, when after an episode of bronchopneumonia she developed features typical of Rett syndrome.

The parents noticed gradual regression of her speech and useful hand movements. The child also developed stereotypic midline hands and lip movements simulating air blowing. Speech and purposeful hand movements were completely lost in 6-8 months. Meaningful play regressed over one-year. Hearing, vision and sleep remained normal. There was no history of seizures prior and during the course of illness. She could still walk but was unable to run/climb stairs. She also has unprovoked bouts of laughter. During examination she spontaneously hyperventilated twice for about 30-40 seconds and had reflex generalized flexor myoclonus to sudden loud sound. Her eye contact and response to pain was very poor. Spontaneous walking and picking things was normal. She did not pay any attention to toys in the playroom and did not obey any command. Tone and deep tendon reflexes were normal. Fingers in both hands were small and tapering. Rest of the examination, including fundi was normal. EEG/ECG and CT scan did not reveal any abnormality. Chromosomal analysis was not carried out.

This child had characteristic features of Rett syndrome including the age at onset, loss of communication and acquired hand skills,

stereotypic hand wringing and washing movements. All her milestones virtually regressed over one year after an episode of bronchopneumonia. Environmental factors like fever has been reported to trigger the onset of RS in genetically predisposed subjects, though there are hardly any reports in the literature. Seizures are one of the supportive criteria but these are overestimated. Non-seizure events like twitching/jerking/head turning/trembling/bruxism/staring/laughing/pupillary dilatation/breath holding and hyperventilation are usually confused with seizures(2). Role of video EEG is very important for diagnosis and to avoid the overprescription and adverse effects of antiepileptic drugs. Myoclonus although reported has not been characterized. Children with RS are prone to sudden death. Prolonged QT interval/low heart rate/hyperventilation and breath holding spells are important components of this syndrome.

Treatment is symptomatic. For seizures lamotrigine is better. Beside seizure control, it improves concentration(3). Children with Rett syndrome have low levels of L-carnitine and vitamin E. These two agents have promising role in the management of Rett syndrome(4). We also prescribed vitamin E and carnitine for four months but the child did not show any improvement. Subsequently the child was put on lamotrigine. After two months her social interaction, visual contact and language improved, but there was no change in her stereotypic hand movements. After six month of treatment she showed significant improvement in her social behavior.

**K .S. Rana,
M.N.G. Nair,**

*Department of Pediatrics,
Armed Forces Medical College,
Pune 411 040, India.
E-mail: kamersinghrana@yahoo.co.in*

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An Outbreak of Mumps in Thiruvananthapuram District

Kerala State has established surveillance of diseases of public health importance using the model of 'district level disease surveillance' (DLDS) developed in the North Arcot district in Tamil Nadu(1). Physicians of both public and private sector health care facilities report 15 notified disease and other diseases at their discretion, through a pre-formatted, self-addressed business reply post card. The card also contains space to enter the basic demographic information and place of residence of all persons with reported illnesses. Space is also provided on the card to enter the date of onset of disease and the date of reporting. The disease reports are received in the office of the District Medical Officer (Health) on all working days. Each card is scrutinized and all data are captured in pre-formed tables, for analysis. Thiruvananthapuram, the southern-most district in Kerala commenced DLDS on January 1, 2002 and the adjacent district of Kollam started it in June 2002.

During January, February and March we received a large number of reports of mumps in Thiruvananthapuram district, clearly indicating the occurrence of an outbreak.

Cases were from all parts of the district. Anecdotally there was information of mumps outbreak in the southern parts of Kollam, but DLDS had not been initiated in that district at this time. The diagnosis made by the pediatricians and physicians was accepted since mumps is relatively easy to diagnose based on clinical features of acute parotitis, unilateral or bilateral. The epidemic curve is presented in *Fig 1*, with the beginning of the outbreak in the last week of January, the peak in the second week of February, and the last cases in the third week of March. The age distribution of cases is presented in *Fig 2*. There was no case below 1 year but there were 52 cases between 1 and 5 years, 29 in boys and 23 in girls. The largest number was in the year-group of 5 to 9, with 98 cases, 54 in boys and 44 in girls. In the age group of 10-14 years there were 25 cases, 15 in boys and 10 in girls. There were very few cases beyond 14 years: one boy and 3 girls 15-19 years old had mumps. Two women in their 20's and one woman in her 30's had mumps and the oldest subject with the disease was a man 42 years old. Age was not recorded for one male and two females. Since mumps is usually a self-limited illness and not associated with fatality, it was not notified for reporting in the DLDS system. Yet, the availability of a disease-reporting system was used by a large number of doctors to report mumps in the