

final outcome of surgical treatment is variable in these children.

## REFERENCES

1. Jones WA. Familial multilocular cystic disease of the jaws. *Am J Cancer* 1933, 17: 946-951.
2. Seward GR, Hankey GT. Cherubism. *Oral Surg* 1957, 10: 952-974.
3. Hammer JE. The demonstration of perivascular collagen deposition in cherubism. *Oral Surg* 1969, 27: 129-141.
4. Topazian RC, Costich ER. Familial fibrous dysplasia of the jaws (Cherubism): Report of a case. *J Oral Surg* 1965, 23: 559-566.
5. Lichtenstein L, Jaffe H. Fibrous dysplasia of the bone. *Arch Pathol* 1942, 33: 777-816.
6. Jones WA. Further observation regarding multilocular cystic disease of the jaws. *Brit J Radiol* 1938, 11: 227-241.
7. Anderson DE, Mc Clendon JL. Cherubism-hereditary fibrous dysplasia of jaw. I-Genetic consideration. *Oral Surg* 1962, 15 (Suppl.): 2: 5-42.
8. Shafer WG, Hine MK, Levy BM. *A Text Book of Oral Pathology*, 4th ed. Philadelphia, W.B. Saunders Company, 1983, pp 694-699.
9. Hamner JE, Ketchman AS. Cherubism: An analysis of treatment. *Cancer* 1969, 23: 1133-1143.
10. Thompson N. Cherubism: Familial fibrous dysplasia of the jaws. *Brit J Plast Surg* 1959, 12: 89-103.
11. Muralidhar M, Shreed S, Vijay Raghavan MR. Monostotic fibrous dysplasia of maxilla. *J Indian Med Assoc* 1987, 85: 273-277.

## The Aicardi Syndrome

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Since, the description of Aicardi syndrome in 1965(1), several cases of this rare entity have been reported(2,3). The characteristics of this syndrome are, its occurrence in females, seizures (often flexion spasms), chorioretinal lacunae, agenesis of corpus callosum, mental subnormality and a variable association of vertebral or other osseous abnormalities. The electroencephalogram has multifocal epileptiform abnormalities with burst suppression, arising independently of two hemispheres or hypsarrhythmia(4). Wadia *et al.* perhaps reported the first and only case from India(5). We report a case of Aicardi syndrome who had asymmetry in EEG.

## Case Report

An eleven month-old girl was referred to the neurological care of G.B. Pant Hospital for the control of her intractable seizures. She started, initially with myoclonic jerks (infantile spasms) since the age of 2 months. She was first in birth order and product of non consanguineous marriage. Both of her parents were healthy. She was born vaginally at 40 weeks of gestation.

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The antenatal, natal and postnatal events were normal. She had received ACTH therapy for infantile spasms. Her spasms had reduced in frequency after ACTH therapy, but never disappeared. She in addition had generalised tonic clonic seizures from 8 months of age. She was on sodium valproate and phenobarbitone in adequate dosages. Her examination, revealed micro-ophthalmia of right eye, chorioretinal lacunae in both eyes and coloboma of left optic nerve. She had generalised hypotonia, normally elicitable deep tendon jerks, bilateral upgoing plantars. Her milestones were of five months old child. The routine hemogram, blood biochemical profile for sugar, electrolytes, calcium, phosphorus, urea, creatinine, acid base was normal. Urinalysis was normal; Results of aminoacid analysis and reducing substances in urine, serum immunoelectrophoresis, cerebrospinal fluid chemistry and cytology were all normal. Serological tests for toxoplasma, rubella, cytomegalovirus, herpes simplex, syphilis were negative. Chest and skull X-rays were normal. She had spina bifida of L5 vertebra on spine X-rays. CT Scan (Head) revealed the absence of corpus callosum and upward extension of 3rd ventricle. MRI examination confirmed the findings of CT Scan and added mild cystic dilatation of 3rd ventricle. EEG (*Fig. 1*) showed dissociated and asymmetrical background activity, being 1 to 3 Hz of 20-30  $\mu$ V on right side and 5 to 6 Hz of 40 to 60  $\mu$ V on left side. The epileptiform spike, sharp wave paroxysm were solely confined to left side. There was no photoparoxysmal response seen.

Her seizures were more or less controlled with sodium valproate, phenobarbitone and clonazepam. But, later clonazepam and phenobarbitone had to be replaced with nitrazepam and carbamazepine

due to uncontrolled seizures. In her followup she had marked mental subnormality. Her EEG, at 3 years of age, showed symmetrical background with occasional runs of asymmetry on two hemispheres. The epileptiform paroxysms were seen bilaterally, more so on right side, but independent on both hemispheres (*Fig. 2*).

### Discussion

The present case had the salient features of Aicardi syndrome: agenesis of corpus callosum, chorioretinal lacunae and infantile spasms. Partial or complete agenesis of corpus callosum may occur alone with sporadic or autosomal inheritance(6). The mode of inheritance of Aicardi syndrome appears to be X-linked dominant with a homozygous male lethality. All cases are sporadic, possibly due to new mutations(7) and female, except one(8). The present case, like previous reports, is female and probably carries a new mutation.

The chorioretinopathy is pathognomic of Aicardi syndrome(2,3,9). It is bilateral posterior in location and consists of white or yellow lacunae of variable size and number, of sharp margins along with little pigmentary change in the adjoining retina. These features differentiates this chorioretinopathy from that of congenital intra-uterine infection, which may be unilateral or bilateral with pigmentary changes and poorly defined margins(9). Other variable ocular anomalies described in this syndrome are colobomas, microphthalmia, retinal detachment, synechiae, etc.(2). The present case had coloboma and microphthalmia in addition to lacunar retinopathy.

The costovertebral anomalies described in the Aicardi syndrome include hemivertebrae, fused vertebrae, fused ribs,

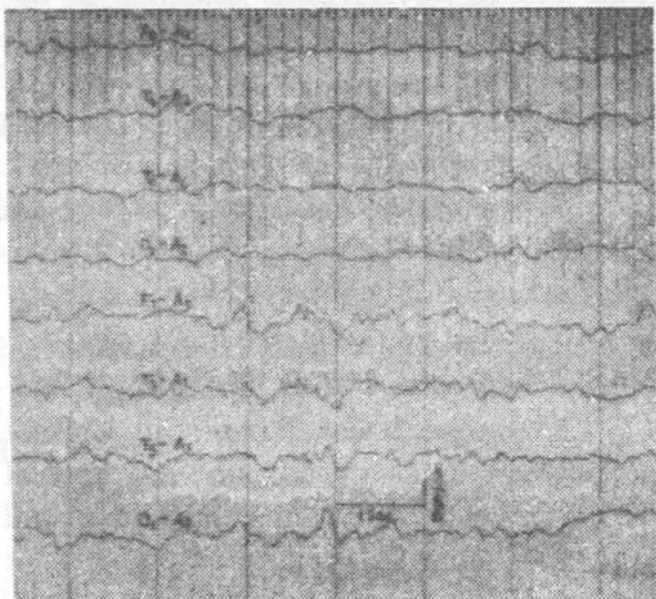


Fig. 1. Showing asynchronous and asymmetrical background activity in both hemispheres.

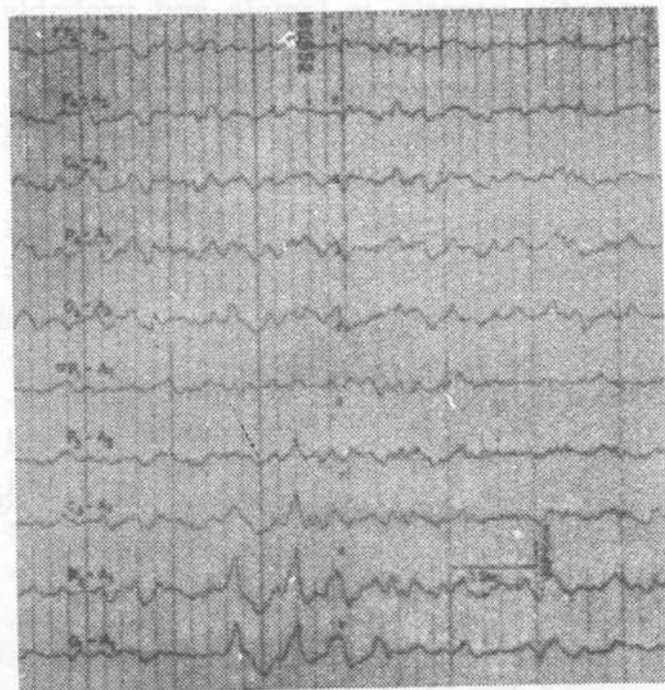


Fig. 2. Showing almost symmetrical background with epileptiform paroxysm in left hemisphere more marked in parietal and occipital leads.

spina bifida occulta, scoliosis(2,3). The present case had spina bifida. The seizures described in Aicardi syndrome as in present case, have always been infantile spasms. Fariello *et al.*(4) believed that an EEG done early in the course of the disease has a specific pattern of asynchronous, multifocal epileptiform abnormalities with burst suppression arising independently from two hemispheres. However, others believe that the EEG pattern is not pathognomic of Aicardi syndrome and may have one or more of following features; burst suppression pattern arising independently from two hemispheres, hypsarrhythmia, diffuse lateralized or bilateral shifting paroxysmal discharges and diffuse lateralised slowing of background activity(2,3). The present case had asymmetric background activity which was independent in the two hemispheres. In the follow up (3 years), EEG was almost symmetrical except brief periods of asymmetry and paroxysmal epileptiform discharges were bilateral, but independent of two hemispheres.

Fariello *et al.*(4) believed that along with anatomical abnormalities of inter-hemispheric connection, there may also be neurophysiological disconnection between cortex and pacemaker. The dissociated EEG in the present case could be an evidence of this anatomical dissociation along with immaturity of cortex, which with age matured to some extent resulting in better developed symmetry of background.

## REFERENCES

1. Aicardi J, Lefebure J, Lerique Koechlin A. A new syndrome: Spasms in flexion, callosal agenesis, ocular abnormalities. *Electroencephalogr Clin Neurophysiol* 1965, 19: 609-610.
2. Denslow GT, Robb RM. Aicardi Syndrome: a report of four cases and

- review of literature. *J Pediatr Ophthalmol Strabismus* 1979, 16: 10-15.
3. Bertoni JM, Von Loh S, Allen RJ. The Aicardi syndrome: Report of 4 cases and review of literature. *Ann Neurol* 1979, 5: 475-482.
  4. Fariello RG, Chun RWM, Doro JM, Buncic JR, Prichard JS. EEG recognition of Aicardi's syndrome. *Arch Neurol* 1977, 34: 563-566.
  5. Wadia RS, Joshi A, Vaidya SD, Manavi AB, Kelkar SR. The Aicardi syndrome. *Neurol India* 1985, 33: 149-154.
  6. Parrish MI, Roessmann U, Levinsohn MW. Agenesis of the corpus callosum. A study of the frequency of associated malformations. *Ann Neurol* 1979, 5: 475-482.
  7. De Jong JGY, Deileman JW, Houben M, *et al.* Agenesis of the corpus callosum, infantile spasms, ocular anomalies (Aicardi's syndrome). *Neurology* 1976, 26: 1152-1158.
  8. Curatolo P, Libutti G, Dallapiccola B. Aicardi syndrome in a male infant. *J Pediatr* 1980, 96: 286-287.
  9. Willis J, Rosman NP. The Aicardi syndrome versus congenital infections: Diagnostic considerations. *J Pediatr* 1980; 96: 235-239.

## BCG Test in Diagnosis of Childhood Tuberculosis

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Tuberculosis is a serious health problem in India with tuberculization occurring to the extent of 80% by the age of 15 years(1). The tuberculin test (TT) is an

accepted intradermal test for international epidemiological surveys but has its own limitations. The observation of the accelerated BCG reaction in children already infected with tuberculosis or tuberculin reactors lead to the development of the BCG test. Inspite of the superiority of the BCG test over the tuberculin test being confirmed by many workers, its use in routine clinical practice is still controversial. Thus this study was undertaken (i) to determine the role of the BCG test in the diagnosis of childhood tuberculosis; (ii) to compare its reliability with TT in the diagnosis of tuberculosis in malnourished children; and (iii) to compare results of TT with BCG test.

### Material and Methods

The study was carried out in the Pediatric OPD of Smt. MT Agarwal Municipal General Hospital, Bombay, over a period of 5 months. One hundred and fifty one children upto the age of 12 years having no previous BCG scar and suspected of having active tuberculosis were subjected simultaneously to BCG test and TT and investigated completely to look for active tubercular lesions. BCG test and TT results were compared in all proved cases of tuberculosis.

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