Infantile spasms syndrome in monozygotic twins. A 7-year follow-up

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The Infantile Spasms Syndrome is a fairly common form of seizures in infancy. Many papers and several books have been published on this syndrome but several aspects are still obscure. In particular, there is some controversy about anticonvulsant treatments and on the question of improvements in mental status. An unusual case of 2 monozygotic twins with this syndrome, both with clinical manifestations appearing within a few hours on the same day, at 6 months has been followed up for 7 years, giving us the opportunity to understand some aspects of the clinical course of the disease and long term treatment.

Key-Words: Infantile spasms – monozygotic twins – ACTH – clonazepan

Although the infantile spasms syndrome is the most frequent form of epilepsy in infancy and many important studies have been published, some points in this disease are still debated. In particular, there is considerable controversy about the efficacy of steroid treatment in the prevention of epileptic seizures and mental retardation [1, 6].

Some years ago we described the unusual case of two monozygotic twins with the infantile spasms syndrome in whom clinical manifestations appeared within a few hours in both patients [3]. A 7-year follow-up of these two boys has enabled us to draw some conclusions regarding the clinical course of the disease and the treatment.

Case reports

These twins were first seen at the age of 8 months. The family history showed no consanguinity or neurological or psychiatric illness except for a distant paternal relative who was said to have had generalised convulsions. Both parents were school teachers and in very good health. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation. The mother first pregnancy had ended in abortion during the second month of gestation.
red at intervals of several seconds with both of 15 to 20 very brief attacks. The episodes recurred several times a day and were considered to be due to colic and no treatment was given. However, the parents became very worried because of the poor psychomotor development in addition to the spasms and so the twins were admitted to the Department of Pediatrics of the University of Catania, where the trouble was promptly diagnosed. With parental agreement a therapeutic trial was decided immediately. In view of the variety of drugs recommended in the literature, two different types of treatment were prescribed [3] twin 1 was given Clonazepam 0.4 mg daily while twin 2 was given ACTH 25 units daily for the first 20 days and every 48 hours for the next 20 days (see fig. 1).

The electroencephalogram was grossly altered with periods of hypersynchronia in both twins just before treatment. 40 days after the first EEG, there was a marked improvement in twin 2 while in twin 1 there was still a considerable amount of irregular slow activity and some discharges. Moreover, although twin 2 had no more spasms at the end of the second week of treatment with ACTH, twin 1 still had spasms, though somewhat less severe.

Since the twin 2 was doing better, twin 1's treatment was changed to ACTH 25 units daily for the first 20 days and every 48 hours for the next 20 days, followed by oral prednisolone for 10 days. The attacks diminished further and then disappeared in the next month. After hormonal treatment phenobarbital was begun (5mg/kg/day), but further bouts of spasms appeared 2 months and 6 months later. At the age of 14 and 18 months other bouts occurred. During the episodes VP (20/mg/kg/day) was added to the PB but the grand mal attacks were still present until the age of 2½ years, when they finally ceased. By the age of 3½ years the anticonvulsant treatment was discontinued. Now this child is attending a normal school with moderate success.

Twin 2, who had been treated first with ACTH for 1½ months followed by maintenance on oral Prednisolone for 10 days with noticeably reduced frequency of the seizures, was put on Clonazepam 0.4 mgs/kg/day. A new course of ACTH at the same dose and period of administration was given when the seizures reappeared at the age of 12 months. The seizures once again ceased and PB and VP were given. After a free interval between the ages of 16 and 24 months, convulsive episodes, myoclonic and GM type, reappeared and still occur. The child is under treatment with PB and VP. His psychomotor development is poor and certainly behind that of his twin.

Laboratory tests in each twin repeatedly yielded