Paediatrica Indonesiana 18 : 109 — 116. March — April 1978.

ORIGINAL ARTICLE

Spasmophilia

by

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Abstract

Sixty eight cases of spasmophilia were studied, consisting of 44 girls and 24 boys, the age ranging from 7 to 12 years. The most prominent symptom was cephalgia. Other symptoms included cramps, fainting attacks, abdominal pains, myalgia, breathing difficulties, palpitation, precordial pain, tremor and learning problems.

Laboratory examination showed that most of them were hypocalcemic with prolonged Qo — Tc interval in the electrocardiogram.

EEG showed a certain pattern of increased thetha waves with irregular background activities and nonspecific changes. Treatment with calcium showed good clinical results. There seems to be no correlation between clinical improvement of symptoms and EMG findings.

Received 22nd. June 1977

Introduction

Not many articles have been published in the literature dealing with spasmophilia as a disease entity; it could be due to the variability of symptoms, or it was considered not a very serious problem.

Spasmophilia or latent chronic tetany is known to be caused by an imbalance of calcium and magnesium both at an intra- or extracellular level, thus producing a state of hyperexcitibility of the neuromuscular system (Bertrand et al., 1966). Clinically it manifests itself by an increased psychic lability, signs of increased mechanical and often electrical irritability of the nerve trunks, as well as various vegetative disturbances (Roth and Nevsimal, 1964). Neuromuscular hyperexcitibility was shown by the decrease in the galvanic and faradic excitation thresholds of the motor nerves, lengthening of the Q — T interval of the electrocardiogram, increased mechanical excitibility, shown by the Chvostek's sign, and typical Electromyographic and Electroencephalographic alterations.

Some authors divided neurogenic tetany from spasmophilia, the latter by the absence of genuine tetanic spasm, while the typical symptoms of hyperexcitibility of the neuromuscular system are present, also the typical EMG pattern of the tetanic syndrome.

Materials and Methods

Material consisted of children with the diagnosis of spasmophilia based on clinical symptoms, EMG, ECG and blood studies; examined at the Outpatient Clinic of the Pediatric Neurology, Department of Child Health, Cipto Mangunkusumo General Hospital in Jakarta, during the period of February 1st, 1975 until January 30th, 1976.

EMG examinations were done and graded according to Djohan Santoso and Jos Utama (1972). Other examinations included Electro-encephalography, Electrocardiography, blood studies, Ophthalmologic examinations to exclude the possibility of visual disturbances, psychiatric and psychologic evaluation if necessary, and in a few cases radiologic bone survey.

Follow up study was done weekly in the first month, and every fortnight thereafter. Treatment was initiated with Ca tablets 3 dd 500 mg. EMG and ECG were reexamined after clinical improvement of symptoms.

Results

There were 68 children diagnosed as having spasmophilia, consisting of 44 girls and 24 boys (Table 1). About half of these children were firstborns (30), and out of these, 16 were the only child in their families, while 16 others were the youngest child.

TABLE 1: *Sex distribution*

Female	44 (64.7%)
Male	24 (35.3%)
Total	68 (100 %)
Siblingship	
First child (only child 16)	30
Youngest child	16
Others	22
Total	68

Age distribution showed the minimal age of 7 (Table 2) and increases with age. The maximal age noted was 12 years, as children above that age were referred to the Department of Internal Medicine.

TABLE 2: *Age distribution*

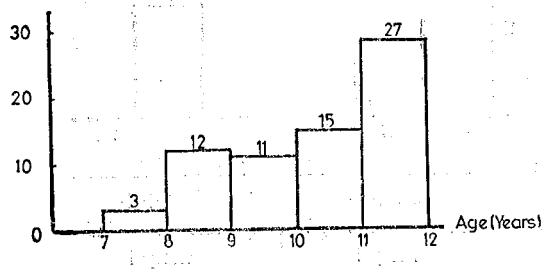


Table 3 showed the presenting symptom that brought the children to the clinic, most were referral cases, a great number of them from the Department of Ophthalmology. The most prominent symptom was cephalgia (43 or 63.1%). The majority of these suffered from attacks or episodes of bilateral headaches, but there were also complaints of migrainous-like attacks. In some children these headaches were so severe that the children had to stay in bed during attacks that may last for hours.

TABLE 3: *Presenting symptoms*

Symptom	Number	%
Cephalgia	43	63.1%
Cramps	7	10.3%
Fainting	5	7.5%
Abdominal pain	3	4.4%
Myalgia	3	4.4%
Breathing difficulties	2	2.9%
Palpitation	1	1.5%
Precordial pain	1	1.5%
Tremor	2	2.9%
Learning problem	1	1.5%
Total	68	100 %

Other symptoms included cramps of the muscles, some had had a carpopedal spasm as well. Fainting attacks were encountered in five children; recurrent abdominal pain in three and other symptoms included myalgia, breathing difficulties, palpitation, precordial pain, tremor and learning problems. Five children who came regularly to the clinic for their convulsive disorders developed symptoms after prolonged anticonvulsant treatment with a minimal of 13 months. On these five children radiological bone survey was done with no evidence of demineralization.

Neurological examinations were outlined in Table 4. Chvostek's sign was

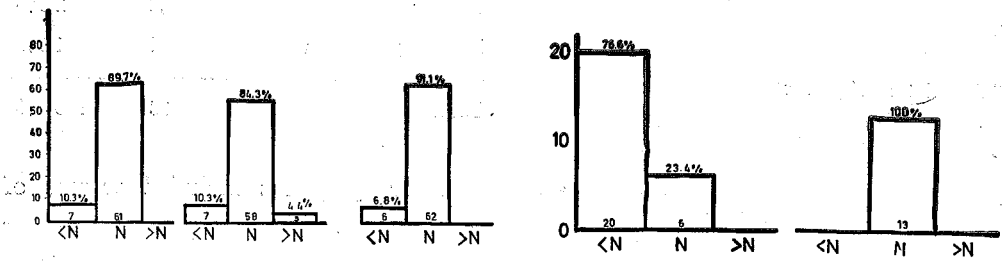
negative in 11 children, one patient had slight bilateral papil edema and 2 showed hyperreflexia of the motor system. Psychiatric evaluation showed character neurosis in two children.

TABLE 4 : *Neurological examinations*

Chvostek's sign	—	11 (16.2%)
	+	43 (63.2%)
	++	14 (20.6%)
	+++	0 (0 %)
Total		68 (100 %)
Bilateral papil edema		1
Hyperreflexia		2

Blood examinations showed mostly normal total calcium levels (89.7%), normal phosphorus (83.3%) and normal alkaline phosphatase levels (Table 5). Serum diffusable ionized calcium levels were examined on 26 children only and serum diffusable ionized magnesium levels on 13 patients; these were due to the limited funds available for laboratory examinations. Out of 26 children, 20 (76.6%) showed a low serum diffusable ionized Ca, while the 13 examined all showed normal serum diffusable ionized Mg levels.

TABLE 5 : *Blood examinations*



Serum total Ca levels. Normal 9 - 12 mg%

Serum phosphorus levels Normal 4 - 6 mg%

Serum Alkaline Phosphatase levels. Normal 10 - 15 K.A.

Serum diffusable Ca ++ Normal 5.6 - 6.7 meq/1

Serum diffusable Mg ++ Normal 1 - 3.5 meq/1

TABLE 6 : *Electrocardiogram (QoTC Inter-val)*

Normal		5 (7.4%)
Borderline (Upper Limit)		2 (2.9%)
Prolonged		
Spontaneous	58	61 (89.7%)
Affer Hyperventilation	3	
T o t a l		68 (100 %)

Qo — Tc interval of the Electrocardiogram was prolonged in 61 (89.7%) even when the Q — T interval was normal (Table 6); 58 spontaneously and 3 after hyperventilation provocation.

Results of the EEG examinations were outlined in Table 7, 8 (11.7%) were normal, 36 (52.5%) were borderline and 24 (35.8%) were abnormal. Included in the borderline cases were EEGs with increased thetha waves or nonspecific changes. Out of the 24 abnormal EEGs, spikes were seen in 3 and 6 records showed occasional spontaneously outburst of high voltaged slow waves.

Follow up study done at the Out Patient Clinic showed that only 44 (64.7%) were regular visitors, while 24 who were not, were regarded as lost to follow up (Table 8). Followed up cases treated with Calcium tablets were divided into 3 groups; group I: If there was disappearance of all clinical symptoms, II : improvement of symptoms without clinical cure, and III : no improvement after 3 months of treatment. In failed cases Mg was added to the treatment.

About 90% (group I and II) showed improvement or disappearance of the presenting symptoms (Table 9). The average duration of treatment was 1.43 months.

TABLE 7 : *Electroencephalographic study*

Normal		8 (11.7%)
Borderline		36 (52.5%)
Increased thetha waves		
Nonspecific changes		
Abnormal		24 (35.8%)
Slow for age	5	
Focal slowing	10	
Spikes	3	
Outburst high voltaged slow waves	6	
Total recorded		68 (100 %)

TABLE 8 : *Follow up study*

Followed up cases	44 (64.7%)
Lost to follow up	24 (35.3%)
Total	68 (100 %)

TABLE 9 : *Folllow up of calcium treated*

Group I	22 (50.0%)
Group II	19 (43.3%)
Group III	3 (6.8%)
Total	44 (100 %)

Re-examination of the EMG was done in the third month on group I and II. Results of the examination as shown in Table 10 were that only 9 out of 41 were improved after disappearance or improvement of symptoms, and none returned to normal.

On 21 patients ECG was also reexamined, 13 returned to normal, 3 showed shortening of the Qo — Tc interval and no improvement was found in 7 children.

TABLE 10 : *EMG re-examination*

Group	No cases	Improved	No improvement	Normal
I	22	6	16	0
II	19	3	16	0
Total	41	9	32	0

Discussion

As was noted before girls were found twice as much as boys in our study and most cases were the eldest or the youngest, while a great number of them were the only child in their family. Most of these parents showed an unusual protectiveness towards these children as was noted during the several interviews. It should be mentioned that for an Indonesian family having only one child is a rarity and the eldest is always regarded as the most important, while the youngest gets the most attention from the others. It seems that psychic lability plays a role in spasmophilia but further study on this matter should be done.

Age distribution showed that the frequency increased with age, 7 years was the minimal age found in our study.

The most prominent symptom cephalgia was also in accordance with the work of Djohan Santoso and Jos Utama (1972), most of these children were referred cases from the Department of Ophthalmology with cephalgia of unknown origin and other possibilities of cephalgia were excluded before the diagnosis of spasmophilia was made.

Very interesting to note were the 5 cases with fainting attacks referred to us from the Department of Cardiology with the possibility of epileptic equivalent and a prolonged Q — T interval in the electrocardiogram. Syncope with prolonged Q — T interval has also been reported occuring in 4 children by Frank and Friedberg in 1972.

About 80% of our children had a prolonged Qo — Tc interval of the electrocardiogram. Prolonged Q — T interval was known to be caused by hypocalcemia. Colletti et al. (1974) in their study on fullterm newborns with hypocalcemia found that prolonged Qo — Tc interval was more frequently than prolonged Q — T interval. The high percentage of prolonged Qo — Tc interval in our study was also in accordance with the high percentage of low levels of diffusable ionized calcium found on

blood examinations, whereas none was found in cases with hypomagnesemia which was also known as a cause of spasmophilia.

Abdominal pain as a symptom of spasmophilia was never mentioned before in other studies. These 3 cases were referred to from the Surgical Department with recurrent abdominal pain simulating and thought to be surgical cases; all showed disappearance of symptoms after calcium treatment.

Five children developed symptoms after prolonged treatment with phenobarbital, in one child in combination with diphenylhydantoin. These children complained of cephalgia which was thought to be a minor sign of their convulsive disorder, but 2 children had experienced cramps of the muscles and carpo-pedal spasm. The EMG examination revealed the typical pattern of spasmophilia, so calcium therapy was initiated with the result of very fast disappearance of symptoms. Radiological bone surveys were normal. Unfortunately due to the limited funds available, diffusable blood ionized calcium and magnesium levels were not examined on these five children.

Hypocalcemia with high alkaline phosphatase levels and signs of rickets were found to occur after prolonged anticonvulsant therapy (Dent et al., 1970, Rickens and Rowe, 1971; Borgsted et al., 1972, Medlinsky, 1974). We could not make any conclusion from these five children although the symptoms disappeared after calcium therapy, as cephalgia could also be a manifestation of their convulsive disorders.

One child was found to have bilateral papil edema which disappeared after 2 weeks of treatment. It can be concluded that it was caused by hypocalcemia, as was also stated by Swash and Roman in 1972 that hypocalcemia was a rare cause of raised intracranial pressure.

Serum diffusable ionized calcium levels were low in 20 of 26 examined cases and in 13 children examinations of diffusable ionized magnesium levels were normal. It can be assumed that cases of hypocalcemia were found rather than hypomagnesemic spasmophilia in our study.

EEG examination showed a high percentage of increased thetha waves with irregular background activities and nonspecific changes (Table 7). Roth and Nevsimal (1964) in their study found that EEG in spasmophilia and neurogenic tetany was mostly normal or slightly changed. Burst of hypersynchronous delta waves during hyperventilation occurred in more than 80% of neurogenic tetany, while these changes were found only in 40,5% of spasmophilia. In our study, outburst of high voltaged slow waves occurred spontaneously without hyper-

ventilation and other abnormalities were more profound. Spikes were also encountered in 3 cases. Swash and Rowan (1972) in their study on a patient with hypoparathyroidism found that spikes also occurred during hypocalcemic states. This is in accordance with the study of hypocalcemia on cats by Schulte and Lohmann (1962).

Follow up study on 44 of 68 cases showed good results with calcium treatment. As can be assumed from laboratory data most of our cases were hypocalcemic. In 3 of the failure group magnesium was initiated with good results in two.

EMG re-examinations on the first and second group showed that only 9 were improved and none returned to normal. Two of these patients came regularly to the clinic after discontinuing of treatment and in one the typical pattern of spasmophilia in the EMG disappeared after 16 months, although symptoms were gone long before. The other one showed only improvement on the last examination. So it seems that there is no correlation between symptoms and EMG pattern in the course of spasmophilia.

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