

/Stamp: DEPARTMENT OF HEALTH OF MOSCOW CITY
SCIENTIFIC & PRACTICAL CENTER OF MEDICAL CARE
FOR CHILDREN WITH CONGENITAL ABNORMALITY
OF CRANIOFACIAL AREA AND INHERENT DISEASES
OF NERVOUS SYSTEM
h.38, Aviatorov str., Moscow, 119620/

CASE RECORD Nr. 622/10

Full name: Sidorin Georgy Vasilievich

Age: 2 years old 7 month

Date of birth: 09.07.2007

Settlement: Moscow city

He stayed at Psychoneurology Department of Scientific & Practical Center **from 18.02.10 till 23.04.10**

Current hospitalization is the first one.

Clinical diagnosis: Cryptogenic localizes (sinistral, frontal) epilepsy, drug-resistant course. Infantile spasms syndrome.

Secondary diagnosis: Minor cardiovascular development abnormalities (abnormal heart chorda). First degree hypertrophy of palatine tonsils.

Complaints at admission: of attacks

Attack type	Frequency	Duration	Time of the day	Registered
Type 1: nods with flexor spasms of upper limbs without loss conscientiousness	6-11 series a day (10-15 attacks per each series)	5-10 minute each series	Often after a sleep and in the afternoon	yes

Treatment at admission: Clonazepam 1,5 mg/d, Suxilep 750 mg/d, Luminal 12, 5 mg/d, Diphenylhydantoin 30 mg/d.

Anamnesis and medical history:

A child was born in normal pregnancy, in time. Pace of development before the attack debut according to age.

13.10.2009 he had an operation - resection of fibrous baby hamartoma of the left hand.

Disease debut: at the age of 2 against the background of absolute health there appeared above mentioned attacks within the waking time, with the frequency of 1-2 per day, duration 5-10 minutes, with gradual acceleration. Since 06.01.10 there have been attacks at night.

Pharmacological anamnesis:

Medicine	Doses mg/d	Terms	Side effects	Concentration (mcg/ml)	Effect
Denakin syrup	450	28.10.09-16.12.09			0%
Clonazepam	2	11.11.09-29.03.10			0%
Suxilep	375	30.11.09-4.03.10			0%
Prednisolone	5	8.12.09-11.01.10			0%
Benzonal	75	15.12.09-20.12.09			0%
Denakin Chrono	500	16.12.09-29.01.10			0%
Lamictal	25	21.12.09-11.01.10			0%

Luminal	12,5	15.01.10-26.02.10		0%
Diphenylhydantoin	30	1.02.10-26.02.10		0%
Denakin chronosphera	500	27.02.10-on-going		>50%
Tonamax	100	17.03.10-17.04.10		<50%
Sabril	2000	9.04.10 – on-going		>75%

Results of previous examinations:

MRT (10.10.09): in the paraventricle parts of the alba of the parietal lobes of both hemispheres – there are unclear areas of myelination retention.

EEG (27.10.09): epileptiform activity with high grade of performance in terms of sets of “acute slow wave” with bitemporal amplitude maximum. Some signals are of regional origin from right posttemporal and left central temporal parts. Separate sets morphologically coincide with DEND.

EEG (1.15.10): there is teta- and delta deceleration registered in frontal – central temporal parts of both spheres. Epiactivity is noticed in terms of several independent foci.

At admission: As to somatic status the condition is satisfactory. Child’s weight is 15,5 kg. Skin and muculent is clear and pink. Heart and lung activity is satisfactory. Physiological functions are normal.

As to neurological status: There are no brain and meningeal symptoms. SN is without focal symptoms, kinesis of limbs is full, tone and power is sufficient. Brisk tendon reflexes, D – S, there are pathological reflexes registered. Coordinational probes are performed satisfactory, sensitivity is not disturbed. Psychic sphere: the child is cooperative, adequate. Speech – separate words, simple phrases.

Examination results:

Indices of blood and urine analysis are within the age norm.

- **Pharmacomonitoring dd. 31.03.10:**
Denakin chronosphere in doses of 500 mg/d: 1 probe: 68 mg/ml, 2 probe: 89 mg/ml.
- **Video-EEG – monitoring during 12 hours with a record of night sleep dd. 22.02.10:**
 - Significant diffuse changes of brain electrobiological activity. Hypsarrhythmia pattern.
 - The sleep is not differentiated in stages. Physiological patterns are not traced.

<i>Characteristic of interictal pathological activity in domination:</i>		<i>State</i>	<i>Localization</i>
Regional deceleration (continuous)	Delta band	SW	C3-T3-T5
Regional epileptiform activity	Spike-deflection Polyspikes	SW	C3-T3-T5
Multiregional epileptiform activity (hypsarrhythmia)	Spike-deflection	SW	
Subicteral patterns	Rhythmic flash wave activity	SW	S>D

Clinical performance:

<i>Attack features</i>	<i>Localization</i>	<i>QTY</i>
Flexor epileptical spasms of different manifestation rate, singular and series	It’s difficult to determine the area of attack start	90%
Flexor epileptical spasms	Sinistral frontal central temporal part	10%

- **Video-EEG- monitoring within 6 hours with a record of night sleep dd. 6.04.10:**
 - Significant diffuse changes of brain electrobiological activity. Hypsarrhythmia pattern. Episodically registered main rhythm corresponds to the age.
 - The sleep is not differentiated in stages. Physiological patterns are not traced.

<i>Characteristic of interictal pathological activity in domination:</i>		<i>State</i>	<i>Localization</i>
Regional deceleration (continuous)	Delta band	SW	C3-T3-T5
Regional epileptiform activity	Spike-deflection Polyspikes	SW	C3-T3-T5
Regional epileptiform activity	Spike-deflection Polyspikes	SW	T4T6
Multiregional epileptiform activity (hypsarrhythmia)	Spike-deflection	SW	
Subicteral patterns	Rhythmic flash wave activity	SW	S>D F3F7

Clinical performance:

<i>Attack features</i>	<i>Localization</i>	<i>QTY</i>
Flexor epileptical spasms of different manifestation rate, singular and series	It's difficult to determine the area of attack start	30%
Flexor epileptical spasms	Sinistral frontal central temporal part	70%

In comparison with the previous examination dd. 22.02.10 there is some improvement in frequency characteristics seen and indices of basic rhythm abundance growth.

- **Brain MRT dd. 23.03.10: there are no pathological processes in the brain.**
- **Psychologist's consultation: Higher mental functions development is slightly lower the age norm.**
- **Speech therapist consultation: speech development delay**
- **Ophthalmologist consultation: OU-development according to the age**
- **Geneticist consultation, Candidate of medical sciences Zhilina S.S.:** in the first place one should exclude the group of genetic leukodystrophy (TMC)-Crabbe, met achromatic leukodystrophies, Tay-Sacks disease.

Treatment applied:

In respect of ineffectiveness, Difenin, Luminal and Clonazepam are canceled. Depakin chronosphere 500 mg/d is introduced, on the back of which the number and duration of attacks has decreased. There also has been Tonamax max 6,5 mg/kg/d introduced, secondary to which the attacks modified – the start with rhythmic low amplitude twitching of the right leg with posterior nod and shoulders uplift, the number of attacks hasn't changed greatly. Sarbil is introduced with the doses increase up to 2000 mg/d (130 mg/kg/d), in this connection there has been a significant decrease in the number of attacks up to 3-4 times per 24-hours. He bears the treatment satisfactory.

Release condition: satisfactory

Dynamics in somatic and neurological statuses: is overall positive in terms of frequency characteristics improvement and indices of basic rhythm abundance growth, significant decrease of attack number, positive psycho verbal development of the child.

Efficiency of antiepileptic therapy (%): decrease of attacks > 75%.

There has been no contact with contagious patients.

Recommendations:

- Denakin chronosphere 250 mg twice per 24-hours with a 12-hour interval,
- Sabril 0,5 2 pills twice per 24hours with a 12-hour interval

There is a future Sabril doses increase up to 150 mg/kg/d possible, provided that there are still attacks but with no side effects.

Antiepileptic medicine should be taken permanently, durably and at the same time without skipping the medicine taking!

Doctor in charge	<i>signed</i>	/Ananjeva T.V./
Chief of the Department	<i>signed</i>	/Yavorskaya M.M./
Head of Epileptology Department	<i>signed</i>	/Ayvazyan S.O./

/Triangular Seal:

**STATE HEALTH CARE INSTITUTION
OF THE DEPARTMENT OF HEALTH OF MOSCOW CITY * PSRN 102776210237
SCIENTIFIC & PRACTICAL CENTER OF MEDICAL CARE FOR CHILDREN WITH CONGENITAL
ABNORMALITY OF CRANIOFACIAL AREA AND INHERENT DISEASES OF NERVOUS SYSTEM
FOR MEDICAL CERTIFICATES/**