Congenital Nystagmus—Report of five cases

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INTRODUCTION

Nystagmus always has meaningful interpretation for clinical evidences. Congenital spontaneous nystagmus give many interesting facts and suggestions to our otoneurologists. So, there are several contributory studies on this disorder referrable. Most of them, however, described the evidences based on the vestibulo-occular findings, but few from the view point of the vestibulo-spinal reflexes. In this paper, we would like to present five cases of congenital nystagmus and its analysis by means of electronystagmography (E N G) and Acceleration-Registrography (A R G).

METHOD

More precise description or methods of static functional A R G and galvanic test will be seen in the previous reports[6)7)]. Nystagmus, either spontaneous or optokinetic, were recorded by the electronystagmography. The deflexions upwards in every curve of E N G represent eye movements to the right and those downwards to the left. Registration of head and body movement in the examination were made by Acceleration-Registrography (A R G). The deflexions upwards in every curves of A R G represent head and body movements to the right and those downwards to the left.

Report of Cases:

Case 1, 25 year-old male, an operator in Telephone Company, was in good health except for persistent eye movement which was indicated by his friend about 10 years ago. He did not note any other symptoms such as nausea, vomit, dizziness, tinnitus and hearing loss. However, it is interesting to note that he can read the books at the right lateral position easier than other position. As shown in the family tree (Fig. I-a), there were other two cases with similar eye movements, otherwise no other notable congenital malformation. No neurological finding was obtained by specialist. Visual acuity was 0.5 on both sides. There was no abnormality in fundi. Physical examination showed normally developed and normally nourished without any notable abnormality in his ears, cardio-vascular system, too. He denied consanguinous marriage in his parents.

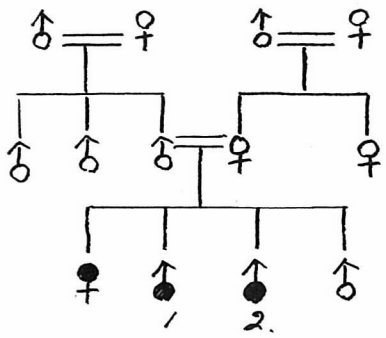

Fig. I-a

Vestibular test: Rotation test (Bárày) and caloric test (0° C, 20cc) showed prolonged duration and increased amplitudes of the nytagmus. Positional test showed no change in direction and amplitude of spontaneous nystagmus. ENG of spontaneous nystagmus as shown in Fig. I-b, when the subject fixed his gaze straight forward, upward or downward, these curves showed that nystagmus was pendular and rhythmic. When gazing laterally to the left, nystagmus increased its amplitude and frequency, and when gazing laterally to the right, diminished remarkable. This suggests that this eye position is the position of rest in this subject.
It is correspond to the fact that he can easily read the book at the right lateral position.
E N G of optokinetic nystagmus showed in Fig. 1-c.
When an optical drum was rotated to the left, the nystagmus was elecited to the left. This was atypical reaction, or in other words, inversion, according to Fisher[1]. However, when the drum was rotated to the right, nystagmus was elicited to the left. This part of the response was normal reaction. Now, we shall give a brief description of the A R G of static functional tests. This test is arranged for detection of latent disturbance of static function of the human body, consisting of

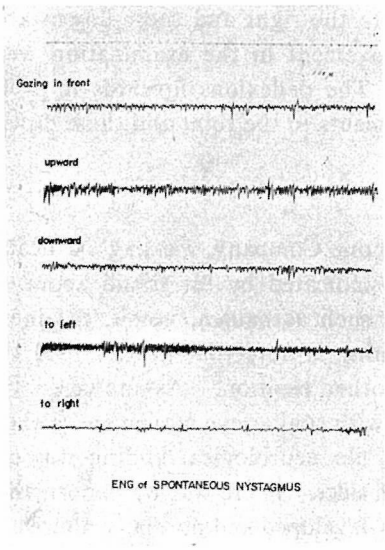

Fig. I-b

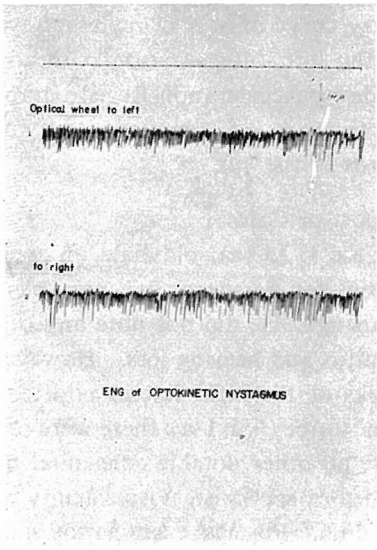

Fig. I-c

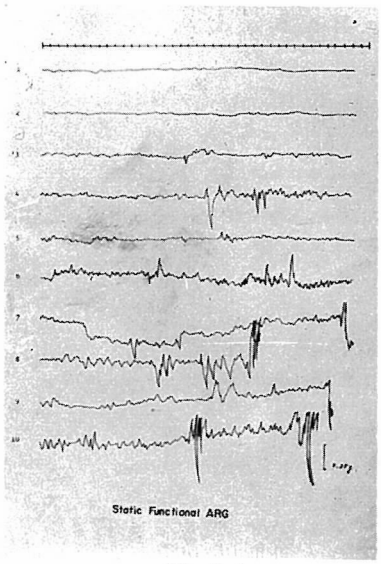

Fig. I-d

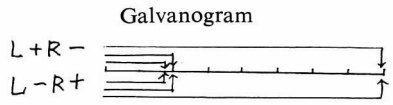

Fig. I-e

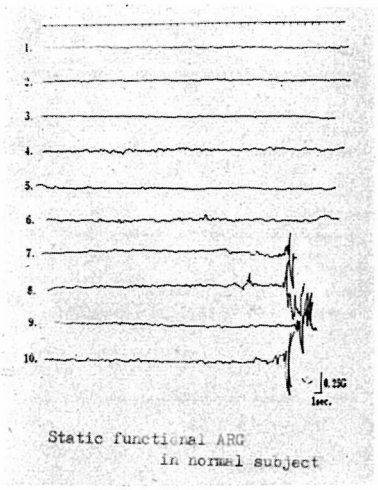

Fig. I-f

A R G recording of the Romberg test, Mann's test and Goniometer test[4)7)]. Each A R G curve is arranged as a graphic pattern. A R G pattern of normal subject showed in Fig. 1-f. The Acceleration Resistrograms of the static functional tests showed in Fig. 1-d. In this pattern, we were able to see slight abnormality with moderate stagger, even though the patient can stand for 40 seconds well. Results of galvanic test showed in Fig. 1-e.
This is a modified galvanic test which we called Galvanogram[6)]. The results were within normal limits (normal and symmetrical excitability).

Case 2, 16 year-old male, a school boy, younger brother of the former (Fig. I-a), had been in heart disease and persistent eye movement since his infantile stage. Howev er, he denied any other symptoms such as nausea, vomit, dizziness, tinnitus and hearing loss. He noted also that he could easily read the book at the right lateral position. There was no history of head trauma, encephalitis or meningitis. Physical examination shows normally nourised male, without any notable cyanosis or clubbing finger. Normal findings were obtained in his ears, nose and throat.
Vestibular test;
Rotation test (Bárány) and caloric test (0°C, 20cc) showed prolonged duration and increased amplitudes of the nystagmus, similar to the former.
Positional test showed no change in direction and amplitude of spontaneoius nystagmus.

E N G of the spontaneous nystagmus and E N G of optokinetic nystagmus, static functional A R G and Galvanogram were in Fig. II–a, b, c, d.

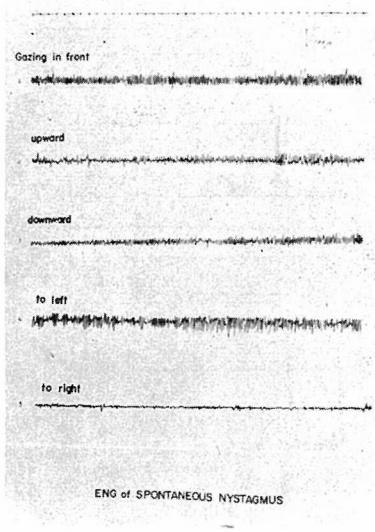

Fig. II–a (Case 2)

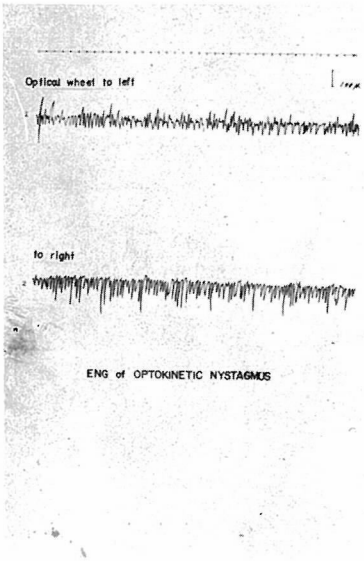

Fig. II–b

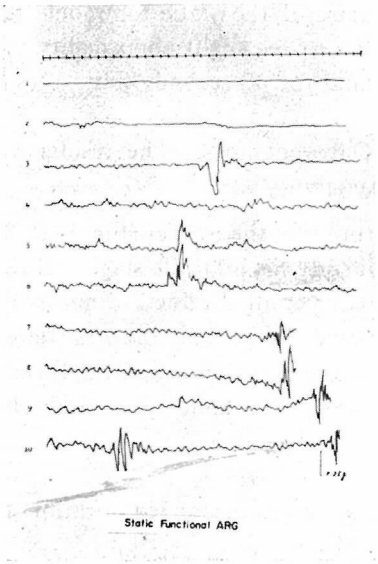

Fig. II–c

Galvanogram

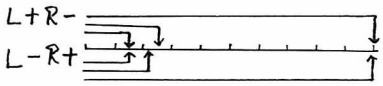

Fig. II–d

When the patient fixed his gaze straight forward, or upward and downward, these curves showed that nystagmus was elicited to the left. When gazing laterally to the left, nystagmus was pendular. But when gazing laterally to the right, nystagmus diminished remarkable. When the optical drum was rotated to the right, nystagmus was elicited to the left, and this is normal reaction. But when the drum was rotated to the left, nystagmus was elicited to the left. This was an atypical reaction, or inverse phenomenon. In this A R G patterns, we are able to find abnormality. Result of the galvanic test was within normal limits.

Case 3, 27 year-old male, bartender, in no notable disease, was referred from ophthalmological clinic with persistent nystagmus which had been noted since his age of 7 years old, without any other symptoms such as tinnitus, hearing loss, dizziness and nausea or vomit.

No head trauma or encephalitis was described. No patient with similar troubles was in his family. He noted for long time that he could easily read the book at the right lateral position. Physical examination showed normally developed and normally nourished, without any notable disease in his ears, cardiovascular system, too. No notable congenital malformation. No neurological finding was obtained.

Vestibular test;

Rotation test and caloric test showed slight prolonged duration and increased amplitudes of the nystagmus. Positional test showed no change in direction and amplitude of the nystagmus. In E N G of the spontaneous nystagmus (Fig. III–a), nystagmus was pendular, when gazing straight forward, and when gazing laterally to the left, upward or downward, nystagmus was elicited to the left, and when gaz-

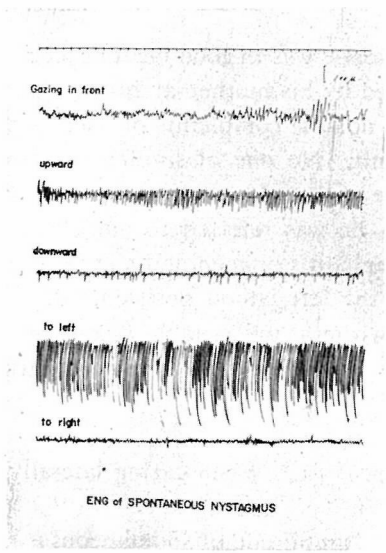

Fig. III–a

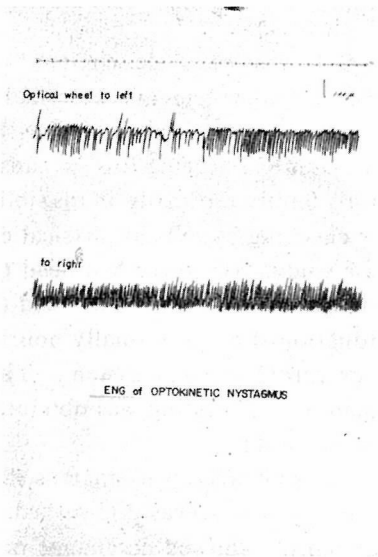

Fig. III–b

ing laterally to the right, nystagmus was diminished.
On turning the optical drum, nystagmus was observed in the same direction as that of the turning. This was an atypical reaction or inverse phenomenon (Fig. III-b).
A R G of static functional test showed in Fig. III-c.
Each curve is nearly normal pattern. In Galvanogram, showed in Fig. III-d, abnormal excitability is demonstrated.

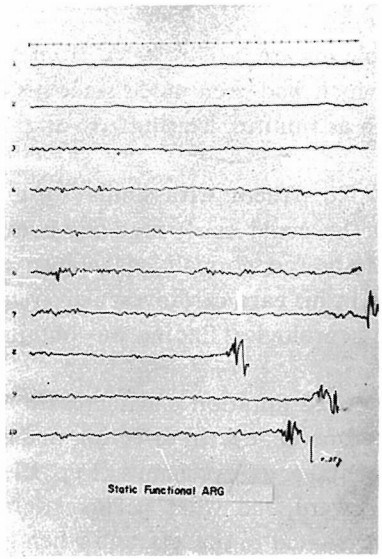

Fig. III-c

Galvanogram

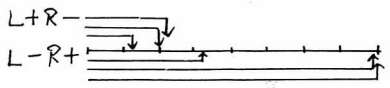

Fig. III-d

Case 4, 41 year-old male, engineer in an iron works, was in good health except for persistent eye movement which had been noticed by his mother at his age of one year, however, no accompanied with any other notable complaints of such as dizziness, tinnitus, hearing loss or nausea and vomit. No one of similar nystagmus is in his family especially in his siblings, but one of them has severely high myopia. After checking by annual physical examination, he was referred to our clinic for precise study. He never had head trauma, encephalitis or meningitis experienced. He noted that he could easily read the book at the left lateral position. Physical examination showed normally nourished male without any notable disease in his ears or cardio-vascular system. There was no notable congenital malformation. No neurological finding was obtained.
Vestibular test ;
E N G of spontaneous nystagmus showed in Fig. IV-a. When gazing laterally to the left, nystagmus was diminished.
Positional test showed no change in direction and amplitude of spontaneous nystagmus.

E N G of optoki-netic nystagmus showed in Fig. IV–b.
On turning the optical drum to the right, nystagmus was elicited to the same direction.

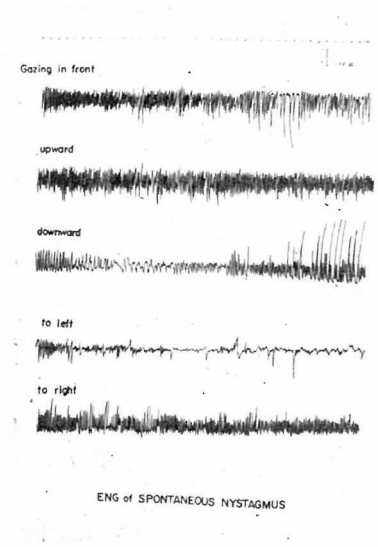

Fig. IV-a

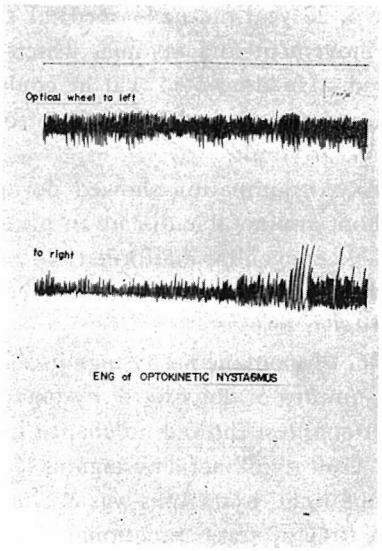

Fig. IV-b

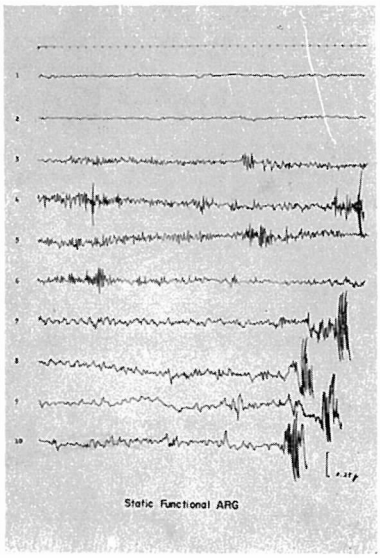

Fig. IV-c

Galvanogram

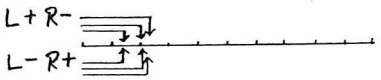

Fig. IV-d

This was atypical reaction.

A R G of static functional test showed in Fig. IV–c.

In Mann's test and Goniometer test, abnormal patterns were demonstrated. Meanwhile Galvanogram showed in Fig. IV–d, normal excitability was noted.

Case 5, 23 year-old male, medical student was fairly good health with persistent eye movement and myopia, which had been noted since the pre-primary school period. He also noted that he could easily read the book at the right lateral position. He never had any other troubles such as tinnitus, hearing loss, dizziness, nausea or vomit.

Physical examination showed normally developed and normally nourished male without any notable disease in his ears or cardio-vascular system. There was no notable congenital malformation. No neurological finding was obtained.

Ophthalmological examination revealed strabisnus and myopia.

Vestibular test;

E N G of spontaneous nystagmus showed in Fig. V–a. When gazing laterally to the left, upward or downward, nystagmus diminished.

Positional test showed no change in direction and amplitude of the nystagmus.

E N G of optokinetic nystagmus showed in Fig. V–b. On turning the optical drum to the right, nystagmus was elicited to the right.

In A R G of static functional test, showed in Fig. V–c, each curve was normal pattern.

Galvanogram revealed abnormal excitability Fig. V–d.

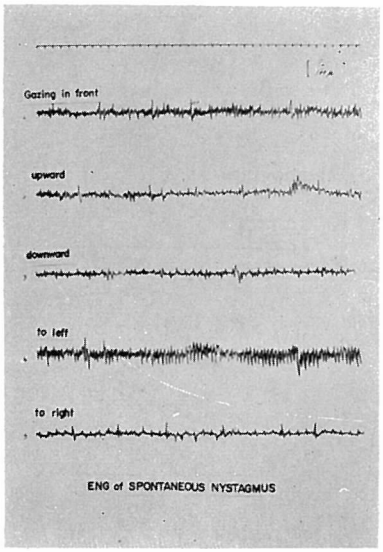

Fig. V-a

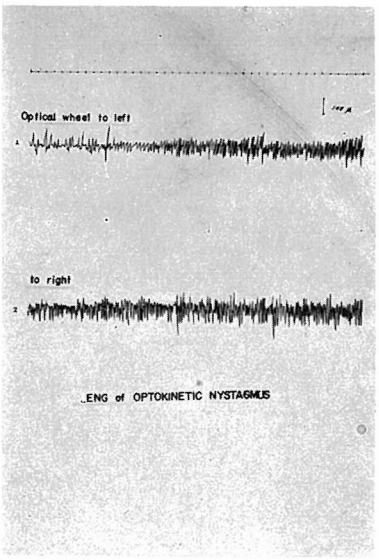

Fig. V-b

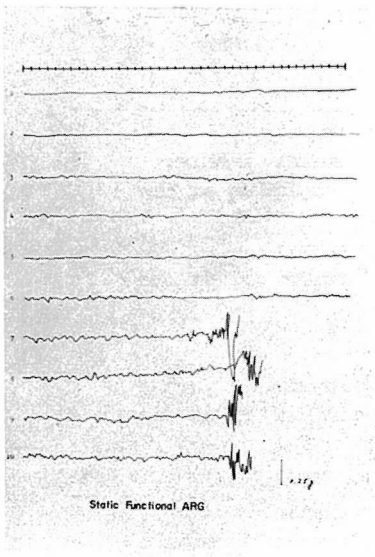

Fig. V-c

Galvanogram

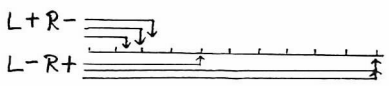

Fig. V-d

CONCLUSION

To summarize the findings and test results of these 5 cases, the following descriptions are made (Fig. VI, Table. I). No patient complained of vertigo subjectively. No one showed positive neurological findings. They all had peculiar eye and head positions, more or less, when looking at the objects. In other words, the position of rest existed. Common and prominent finding was that the nystagmus is pendular, rhythmic, persistent and spontaneous. This pendular nystagmus changed to jerky as lateral gazing.

Another finding was an eccentric position of rest. Four of the five cases in our series have the position of rest on the right side. One has it on the left side.

Optokinetic nystagmus:

According to Fisher[1], all five cases were elicited as "atypical reaction" or in other words, "inversion phenomenon". Furthermore, this inverted optokinetic nystagmus was observed when the optical drum is rotated to the left. Four of the five cases were inverted to the left, and one to the right.

A R G of static functional test:

Some of our cases with congenital nystagmus showed mild to moderate stagger, and unusual pattern which never had seen in normal, in spite of normal duration of standing upright during the tests. In the results of galvanogram, some of this series showed abnormal responses to galvanic stimulation.

Two of the five cases showed normal and symmetrical excitability. Three of them

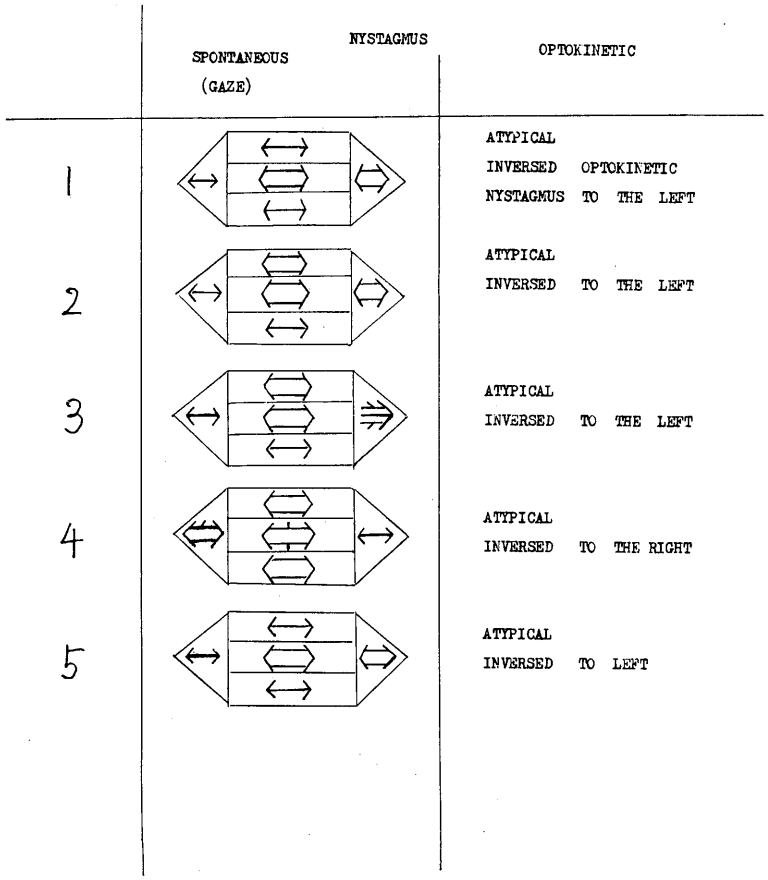

Fig. VI

Table. 1 Summary of VESTIBULO-SPINAL REFLEXES of fiue cases.

Case No.	STATICFUNCTIONAL A R G	GALVANO—A R G
1	MODERATE STAGGER NORMAL DURATION	NORMAL SYMMETRICAL
2	MILD STAGGER NORMAL DURATION	NORNAL SYMMETRICAL
3	NORMAL	ABNORMAL, EXCITABLE ASYMMETRICAL (R>L)
4	MODERATE STAGGER NORMAL DURATION	ABNORMAL, EXCITABLE SYMMETRICAL
5	NORMAL	ABNORMAL, EXCITABLE ASYMMETRICAL (R>L)

demonstrated abnormal excitability. Two of the three showed a symmetrical responses. The right side was more excitable than the left side.

COMMENT

The cause of congenital spontaneous nystagmus is unknown[2),3)]. The findings presensed suggest that the origin is not labyrinthine but ocular-central. It is thought that the brain stem or midbrain may be responsible[3),4),5)]. Fourthermore, the location may be at and/or nearly the superior colliculus (oculomoter nuclei).

SUMMARY

Five cases of congenital spontaneous nystagmus were presented, which showed pendular, rhythmic and persistent eye movement; and atypical reaction or inversion to optokinetic stimulation.

REFERENCES

1) Fisher, J. J.: *The labyrinth, physiology and functional tests*, Grune & Stratton, New York, 1956.
2) Forssman, B.: A study of congenital nystagmus, *Acta Otolaryng.*, **57** : 427, 1964.
3) Anderson, J. R.: Causes and treatment of congenital eccentric nystagmus, *Brit. J. Ophthal.*, **3715** : 267, 1963.
4) Tsujikawa, O. et al: Congenital familial nystagmus, *Otolaryng.* (Tokyo), **37** : 435, 1965.
5) Suzuki, J,: A stady on pendular nystagmus, —pendular nystagmus; Its contribution to the understanding of nystagmus mechanism, *Acta Oto-laryng.*, **53** : 381, 1960.
6) Sekitani, T.: Galvanic test—Acceleration Registrogram of head movement induced by galvanic stimulation, and Galvanogram (Sekitani), (Japanese with English abstract), *Jap, Jour, Otol*, (Tokyo), **68** (8) : 996, 1965.
7) Tsujikawa, O.: Human Equilibrium in Standing position (Japanese with English abstract), *Jap. Jour Otol.* **68** (8), 1016. 1965.

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