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IDIOPATHIC PROGRESSIVE VESTIBULAR DEGENERATION IN A YOUNG MAN:  
LOSS OF VESTIBULAR SERVATION NOT THE BASIS FOR DETECTION

Ashton Graybiel, Curtis R. Smith, Fred E. Guedry, Jr., Earl F. Miller II,  
Alfred R. Fregly, and D. Bryant Cramer



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NAVAL AEROSPACE MEDICAL RESEARCH LABORATORY  
PENSACOLA, FLORIDA

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## SUMMARY PAGE

### THE PROBLEM

This report presents the case of a young man, TI, who experienced episodes of dizziness and demonstrated the rare combination of bilateral loss of nonacoustic function with retention of normal hearing. Persistent dizzy spells first appeared in childhood and were usually mild, but, on rare occasions, he was handicapped by brief severe attacks of vertigo.

### FINDINGS

TI was nearly 26 years of age before these episodes were shown to be of vestibular origin; yet, during the preceding years he never complained of symptoms attributable to loss of function, as distinct from perturbed vestibular function. At that time residual function was substantial in the otolith organs but minimal and nil in the vertical and horizontal canals, respectively. When last seen at age 29, even otolith function was lost; this was associated with the gradual disappearance of dizzy spells. The diagnosis, idiopathic progressive vestibular degeneration, was made by exclusion. The notable conclusion to be drawn from this patient was not the fact that he had dizzy spells of vestibular origin, but the fact that loss of vestibular function per se was not incompatible with leading a life regarded by himself, his family, and friends as normal.

## INTRODUCTION

Less is known concerning normal functioning than abnormal functioning of the vestibular system in man. These otolithic and canalicular systems serve mainly as "silent participants" in motor functions, and it is exceedingly difficult to elucidate this participatory role as it affects man's behavior under natural circumstances. A classical experimental approach in attempting to evaluate this role involves the use of human (1-3) or animal (4-6) subjects with bilateral loss of canalicular and otolithic functions. Experiments on animals have shortcomings, partly because of technical difficulties but mainly because the findings are not directly applicable to man (7). The identification of human subjects with bilateral loss of vestibular function has been accomplished either in the clinic or by screening groups of deaf but otherwise normal persons. But the behavioral findings on patients may be suspect, and findings on deaf subjects may be difficult to interpret because of the great differences between persons who hear and those who do not. Although the identification of persons with normal hearing and loss of vestibular function would provide ideal subjects, the screening procedure entails a major undertaking because of the extreme rarity of such cases in a general population. Few cases of isolated loss of vestibular function in otherwise normal young persons have been thoroughly evaluated, hence our interest in the case to be reported.

## CASE REPORT

Toward the end of May 1966, a pleasant-mannered married man, who had recently been inducted into the Army as a private, applied for a commission on the grounds that he was a registered public health sanitarian with a college degree. In filling out one of the forms he answered "yes" to the query, "Have you ever been dizzy?", which initiated a chain of events leading to his entry into the Brooke Army Hospital and subsequent referral to our vestibular laboratory. For the first time T1, then nearly 26 years of age, learned that his organs of equilibrium were not functioning normally and that this dysfunction was connected with his dizzy spells, previously ascribed to "nerves."

An attempt will be made in Section I to provide an account of T1's symptoms and life activities prior to diagnosis, avoiding retrospective interpretation. Section II will deal with the studies conducted in June and July 1966, and Section III will consist of a brief follow-up report.

### I — PREDIAGNOSIS PERIOD

Information regarding T1's early life was supplied by his mother. T1 was born September 16, 1940, the eldest of four sons. Forceps were used in delivery, and he had a "black eye with a swollen face" when first seen. He sat at 5 months, crept at 6 months, stood at 8 months, and walked well at 10-1/2 months; his brothers did not walk until they were about a year old. When 8 months of age, T1 fell downstairs without apparent ill effects. He was severely ill with measles when 4 years of age, and mildly ill with scarlet fever when 6 years of age.

### Elementary School

Between 10 and 15 years of age TI was afflicted with "numerous boils" and was seen by a physician for repeated attacks of "ear infection with drainage." During this period TI experienced occasional "dizzy spells" but recalled few specifically. When about 10 years of age, TI recalled telling his father one morning that he was dizzy, and when asked if he was constipated, became embarrassed and "let the subject drop." Another remembered attack occurred 2 years later during a Little League baseball game. TI stated, "This one stands out because I was on the field at the time. I was able to remain standing, and it passed quickly." He also recalled having dizzy spells in bed and "feeling pleased" that he was lying down.

While in elementary school TI became much interested in sports. He was on the Little League baseball team, was active in the gymnasium, and participated in water sports.

### Secondary School

During this period TI experienced occasional but persistent dizzy spells of varying severity, mostly mild. One occurred while he was in the hallway between classes. He said he had to hang his head down until it was over.

He continued to play baseball, and a "scout" told him he was capable of playing on a college team. He played football, 1 year on the junior and 2 years on the senior varsity teams. Moreover, he played ice hockey (1 year on an organized team), was successful at tumbling and on the trampoline, and swam underwater without face mask and snorkel.

Throughout these school periods TI enjoyed riding carnival devices of different sorts and never experienced motion sickness; he estimated the number of rides in the "hundreds."

### College

Throughout college TI experienced mild spells as often as two or three times a week, although there were periods as long as 3 months when he was free from attacks. He did not recall a severe episode of dizziness while in college. TI failed to make the freshman baseball team but played handball, tennis, and golf.

### Post College

TI graduated in 1963 and found employment as a sanitarian, which involved much driving. He continued to experience occasional dizzy spells but was not afraid of dizziness while driving because usually he was able to "suppress" the symptoms; on one occasion, however, he pulled off to the side of the road.

In July 1965, TI and his wife spent a 3-week vacation at the home of her parents where he experienced frequent episodes of dizziness. Soon thereafter he "was bothered by brief 'flashes' of dizziness every time my ears 'popped' as I changed altitude" (in an elevator or when driving over hills). Rarely, TI suffered what he termed a "major attack." Such attacks were characterized by an abrupt onset with objects "whirling about me to the left." The duration was 2 or 3 minutes, and typical symptoms were nausea, dizziness, and double vision. Movement of the head would aggravate the symptoms. Tinnitus and deafness were never experienced. An attack might end abruptly or be followed by a headache "resembling an eyestrain headache." More details were not obtainable. These experiences prompted his first visit to a physician; a "routine ear, nose, and throat examination was carried out, and some x-rays were taken." The physician said he would report by phone if any abnormality was found; a call was not received.

In January 1966, he experienced a major attack while inspecting a restaurant. Again he sought the aid of a physician and was told that his problem was "nerves."

On February 8, 1966, he was inducted into the Army and in May applied for the commission mentioned earlier.

#### Comment

During the period under review TI summarized his problem in these words:

I have been fairly active in sports throughout my school years, and I have never hesitated to do anything for fear of dizziness or loss of balance. I have led a fairly active life and only occasionally have I been incapacitated by my dizziness. I have learned to live with and around these spells.

TI, his family, and the physicians who examined him considered that he was a person with occasional dizzy spells, but otherwise normal.

## II — MEDICAL EVALUATION

The medical studies conducted at the Brooke Army Hospital and the Naval Aerospace Medical Institute covered a period of 2 months. All of the findings have been combined and will be presented and discussed in the nature of a final summary.

#### Additional History

TI was questioned at length regarding his past history in the light of the diagnostic findings. Only in retrospect, and aided by direct questions, did it become apparent to TI that, during periods when he was free from dizzy spells, he might have been handicapped. He "admitted" that in getting about the house in darkness he used contact cues,

but there was no way of determining whether, for him, this constituted a decrement in performance. With regard to water sports, he recalled that he was "never an accomplished springboard diver," and, "while playing college baseball, I noticed a lack of the balance I had had in the past and a clumsiness that I had not noticed before. These developments hindered my play so that I was dropped from the team." In the prediagnosis period it was reasonable to ascribe these possible handicaps to lack of aptitude. Even in retrospect it is difficult to determine how much TI was handicapped in sports due to vestibular disease.

With regard to the family history, one brother in college wrote that he had "brief dizzy spells" which he thought were similar to TI's and that they were more frequent "when I was younger." They "come and go quickly," lasting a few seconds; "sometimes I have to sit down and put my head between my knees." The only other member of TI's family complaining of dizziness was his mother's sister who experienced this symptom in connection with migraine headaches. His social history left nothing to be desired; he neither smoked nor used drugs and drinking was limited to "an occasional beer."

The record TI kept revealed a relatively large number of episodes which he ascribed to: 1) the strain and fatigue while the vestibular studies were in progress, 2) the fact that he was "looking for these episodes to occur," and 3) "I do feel that the tests (in Pensacola) did tend to stimulate their occurrence." There were one major and two minor episodes of vertigo and at least 5 days when "light-headedness" was experienced for long periods.

The major attack of vertigo occurred on the way to Pensacola. He was sitting across from his wife in a restaurant when he experienced true vertigo for about 1 minute, and his wife noted "irregular horizontal rapid beating of the left eye with little or no right eye motion." At this time there was stomach awareness, and the vertigo was followed by "lightheadedness" lasting about 15 minutes. He did not experience tinnitus or hearing loss.

#### General Medical Evaluation

Except for symptoms referable to the labyrinth, TI had no complaints. Systematic inquiry covering the various regions and functions of the body did not reveal any significant departure from the normal.

Physical examination revealed a well-developed, attractive-appearing young man. He was well poised, alert, and wholly cooperative. Examination of the nose, larynx, neck, chest, and abdomen revealed no definite abnormality. The heart was not enlarged; no murmurs were heard; the pulse rate was 84; and the blood pressure 128/96 mm Hg. The lungs were clear; the thorax resonant; and the roentgenograms of the chest revealed no abnormality of heart or lungs. The findings obtained on urinalysis, blood morphology, and routine chemical tests were within normal limits. The VDRL flocculation test for syphilis was nonreactive.

The neurological consultant stated:

There was nystagmus on conjugate deviation of the eyes to the left with rapid component to the direction of gaze which is sustained. There is similar nystagmus to the right with rapid component to the direction of gaze which is not sustained. With positional tests, the patient, suddenly lying on his back rotating his head either to the right or the left, nystagmus was prominently accentuated without latency. This does not seem to fatigue although vertigo was noted only transiently on the first attempt. The only other significant abnormality was with his eyes closed, the patient was totally unable to walk tandem. He staggered and was grossly ataxic.

Roentgenograms of the skull revealed rather prominent vascular markings; the sella turcica was normal and there were no intracranial calcifications. The electroencephalograms obtained before and after hyperventilation did not reveal any changes outside the normal range.

The general examination of the eyes and the consultant's findings revealed no abnormality; the examination included funduscopy, tests of pupillary and optokinetic reflexes, visual fields, and visual acuity (with contact lenses).

The otoscopic examination and a "mastoid series" (roentgenograms) revealed no definite abnormality. Examination of cochlear function revealed normal hearing (Figure 1); 100% speech discrimination bilaterally; SISI 0%; Bekesy Type 1.

### The Vestibular Organs

In describing the findings, often they will be compared with those obtained on an elite group of deaf persons with bilateral labyrinthine defects (L-D subjects) whose pathological states had stabilized (3). Difficulties both in conducting certain tests and in interpreting the findings were encountered and were attributed either to activity of pathologic processes or to the consequent state of vestibular disequilibrium such injury engendered. Even when TI was not experiencing an episode of dizziness, there might be signs of vestibular disturbance; i.e., positional nystagmus or spontaneous nystagmus. At all times there was gaze nystagmus.

The Postural Equilibrium Test Battery. These tests have a limitation in the sense that many systems in addition to the vestibular reflexes are challenged, but a great advantage in the fact that they test natural behavioral mechanisms.

The test battery, described elsewhere in detail (8) and comprising five individual items, requires the patient to stand or walk in the stringent position of body erect, arms folded against chest, wearing shoes with flat heels and leather soles. The tests were administered in the following sequence:

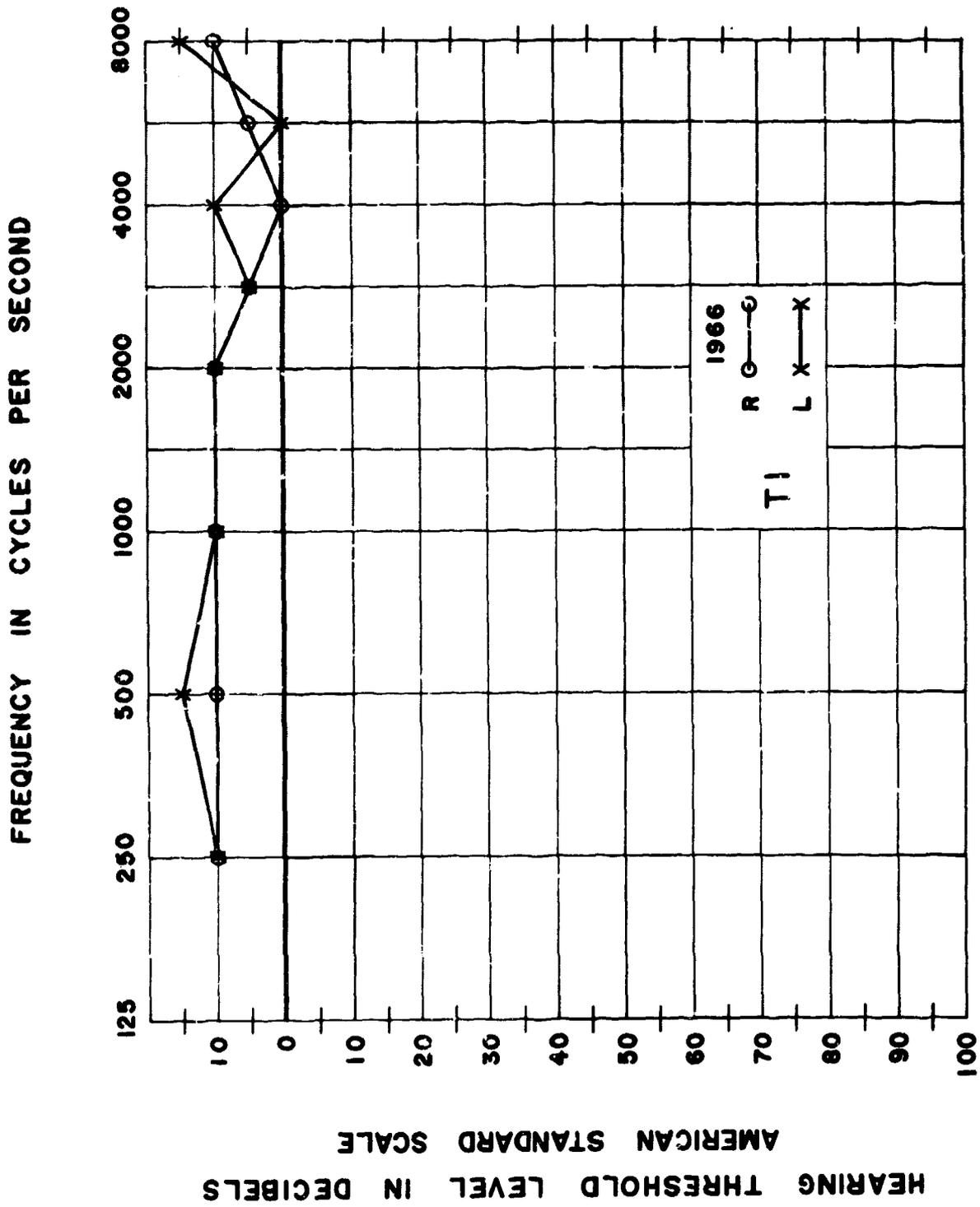


Figure 1  
Hearing threshold level of T1 on medical evaluation in 1966.

1. Sharpened Romberg (SR): Stand on floor, eyes closed, feet in heel-to-toe position. Maximum score 240 seconds.
2. Rail Test Battery (short version)
  - a. Walk eyes open (E/O): Walk heel-to-toe, eyes open, on 3/4-inch by 8-foot rail. Maximum score 15 steps.
  - b. Stand eyes open (E/O): Stand heel-to-toe, eyes open, on the 3/4-inch wide rail. Maximum score 180 seconds.
  - c. Stand eyes closed (E/C): Stand eyes closed on 2 1/4-inch by 30-inch rail. Maximum score 180 seconds.
3. Stand on leg eyes closed (SOLEC): First right leg (SOLEC-R), then left leg (SOLEC-L). Maximum score 150 seconds.
4. Walk a straight 12-foot line on the floor heel-to-toe, eyes closed (WALEC). Maximum score zero (inches deviation from line).
5. Walk on floor eyes closed (WOFEC). Administered simultaneously with WALEC. Maximum score 30 steps.

Sharpened Romberg scores for TI were far below those of normal subjects, and there was little or no evidence of learning. Walking the rail with eyes open resulted in poor scores on the first three occasions but, thereafter, scores were within the normal range. Standing on the rail either eyes open or closed yielded poor scores with little or no evidence of learning. Standing on one leg eyes closed resulted in extremely poor scores and no evidence of improvement. Attempting to walk a straight line eyes closed resulted in variable scores; nine of twelve attempts were extremely poor and three were in the lower normal range. The same test scored in terms of number of steps accomplished yielded scores usually below the normal range.

TI's performance differed from that of a group of L-D subjects (9) in two respects. First, there was greater variability in scores made on certain tests, possibly reflecting the fact that, in contrast to the L-D group, TI suffered spontaneous vestibular disturbances, indicating an active rather than a quiescent process. Variations in this activity, even at the subclinical level, might have influenced ataxia test scores. Second, in those tests in which TI's scores were higher than the means of the L-D group, his overall performance was no better than that of ZA, the top performer in the L-D group. Our findings in subjects with partial loss of function (10) suggest that postural equilibrium is more dependent on canalicular than on otolithic function.

**Otolith Function.** Three tests were used: ocular counterrolling, the oculogravic illusion, and rotation about an Earth-horizontal axis. Counterrolling of the eyes is associated with body tilt away from the upright in the frontal plane (11). A natural behavioral reflex is exploited in this test, although the subject is positioned passively. Under rigid test conditions it has the unrivaled advantage of depending almost entirely on the release of an otolithic reflex. The ocular counterrolling index is defined as one-half the sum of the maximum right and left ocular torsion expressed in minutes of arc. The values obtained in twelve tests conducted on five different days ranged from 66 to 149 (mean index 110). This great variation in index values is in itself unusual and probably abnormal. The highest value, 149, reaches well into the lower end of the normal range, while the lowest value is typical of that obtained on L-D subjects. The results indicate some residual otolith function, and the great variation in values may be indicative of an active underlying pathologic process.

The oculogravic illusion test (12) is conducted on a centrifuge, and the subject's task is to set a dim line of light to the perceived horizontal, while under the influence of a known change in direction of the gravito-inertial horizontal with reference to his body. Water immersion studies (13) have demonstrated that the major contribution to the illusory motion in normal subjects has its origin in the otolith apparatus and, in the L-D subjects, in nonvestibular proprioceptive mechanisms. In Figure 2 are compared the settings made by TI and those made by a group of normal and of L-D subjects, using the same method. With trials randomized insofar as possible, and after stability was reached at each level of centripetal force, the subject set the line to the perceived horizontal after it had been offset by the experimenter. Each point on the graph represents the mean of ten settings. It is noteworthy that the variance of the settings was similar for TI and the normal group and far less than for the L-D group. Although TI's responses were below typical normal values, they were within the normal range.

The third test conducted appears to be a practical test of the dynamics of otolith function. TI was rotated on an electric posture table at constant speed about an Earth-horizontal axis at 10 rpm for 90 seconds several times on two separate days. Under these circumstances, normal persons exhibit a persistent, continuous, unidirectional nystagmus that cannot be attributed to angular acceleration. Subjects without canal function and with minimal or no otolith function either do not exhibit this response or the response (nystagmus) is abnormal and weak (14).

On the first day, TI exhibited a strong direction-reversing horizontal nystagmus during constant speed rotation. His response was not normal for either direction of rotation because there was a position-keyed reversal of nystagmus, but TI also differed from men without labyrinthine function in that a clear response was present.

On the second day, TI was experiencing dizziness prior to testing, and he had a strong left-beating spontaneous nystagmus. On this day his responses during counter-clockwise rotation were comparable to those of normal subjects, whereas his response during clockwise rotation was again direction reversing. The results of this test indicate that otolith function was present but abnormal. The fact that it changed from one day to

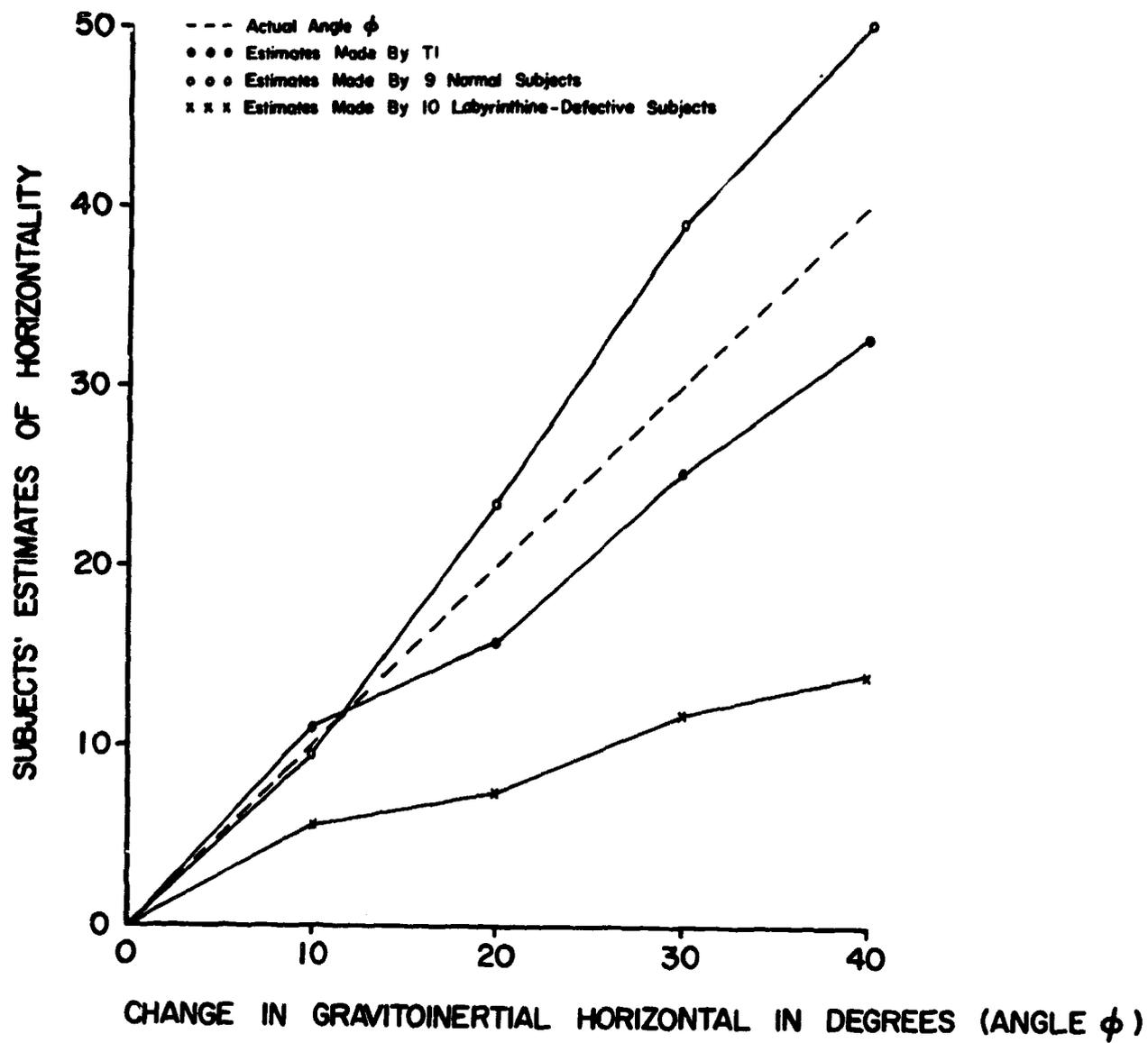


Figure 2

Settings of a dimly illuminated line (in darkness) to the perceived horizontal when a subject was exposed to four changes in direction of the gravito-inertial horizontal with respect to himself. Each point on each curve represents the mean of ten settings.

the next indicates that this abnormal condition of the otolith response system was in a state of change, probably due to an active degenerative process.

In summary, the measurements obtained on all tests of otolith function suggest that TI had reduced function but that it was nonetheless sufficient to contribute to behavioral mechanisms.

Caloric Tests. Thermal stimulation was used on five experimental days. On day one, a modified Hallpike test was conducted (eyes closed, EOG recording), and a left-beating nystagmus was observed in the right- and left-lateral as well as the supine positions. This response, however, was not affected by the cold or warm irrigation. On day two, the Hallpike test was repeated, with similar results except that the spontaneous nystagmus was right beating; subsequent irrigation with ice water (L and R) did not alter the spontaneous nystagmus. On day three, right-beating spontaneous nystagmus was not affected by irrigating temperatures of  $44^{\circ}\text{C}$  and approximately  $3^{\circ}\text{C}$ . On day four, 95 minutes after TI drank 150 cc of 100-proof vodka in orange juice, at which time he did not manifest spontaneous nystagmus, there was no response to irrigation with ice water. On day five, TI was given 25 grains of aspirin to minimize the pain incidental to irrigations of 3 minutes' duration with water around  $3^{\circ}\text{C}$ . Right-beating spontaneous nystagmus was observed off and on. During irrigation on the right side, and in the absence of spontaneous nystagmus, a few deflections suggestive of left-beating nystagmus appeared. During irrigation on the left side, right-beating nystagmus appeared, similar to the spontaneous nystagmus.

Angular Acceleration Tests. Attempts were made to determine threshold responses to angular accelerations. Rotation about the vertical axis with head positioned to stimulate the horizontal canals was carried out on four different days, using a Stille chair. With nystagmus used as an indicator, attempts were made to establish a threshold of response, to elicit a response at maximum stimulation ( $15 \text{ deg/sec}^2$ ) and after sudden stops following clockwise or counterclockwise rotation. On one occasion when left-beating spontaneous nystagmus was present, angular acceleration appeared to have some effect on the spontaneous nystagmus in one trial, a possible effect in one other, and no effect in three trials. On three other days angular acceleration either failed to elicit nystagmus or influence spontaneous nystagmus if present. Moreover, no definite sensation of rotation was elicited, and, in additional trials, the oculogyral illusion (15) was not perceived. It was concluded that the horizontal canals did not respond under these stimulus conditions.

TI was exposed to angular accelerations on the Stille chair and with appropriate head positions in an attempt to stimulate the vertical canals. A weak vertical nystagmus downward was produced on two occasions, in one case with the left and in the other with right ear down, by stimuli that would produce the same direction of beat in normal subjects. Reversing the direction of these stimuli did not produce the expected nystagmic response but seemed to cancel a weak spontaneous nystagmus downward. Coriolis accelerations were generated by directing TI to tilt his head while rotating at 30 rpm. This resulted in vertical nystagmus in the expected direction, but the responses were weak compared with those expected from normal subjects.

On two occasions separated by five days, T1, in the slow rotation room, was required to make single head-body motions (front, back, right, left) with eyes open and eyes closed while rotating either at 10 rpm or 15 rpm. On the first occasion at both rotational velocities when moving forward and backward he felt as if his body described a loop; this was less pronounced moving backward and not experienced moving sideways. This so-called "giant hand" effect is experienced both by normal and L-D subjects, indicating a large nonvestibular component. Rotating at 15 rpm with eyes open, he noticed a little "eyestrain" at the beginning of each movement, and "things looked fuzzy for half a second" at the start and end of each movement. Definite dizziness was not experienced. On the second occasion the findings were similar except for his experiencing dizziness or a "floating sensation" on return of the head to the upright after a forward movement. On both occasions dizziness was much less than might be expected from normal persons.

Interpretation of the findings in terms of vertical canal function is made difficult by inconsistent findings, possible positional effects, and the fact that it is impossible to stimulate the vertical canals by angular or Coriolis accelerations without, at the same time, stimulating the otolith organs in an unusual manner. It was concluded that on the basis of these tests, the possibility of some residual function of the vertical canals could not be excluded.

Provocative Tests. The so-called Dial Test is one means of measuring susceptibility to motion sickness (16). The subject, seated in the slow rotation room, is required to move his head (body) through five different arcs out of the plane of the room's rotation, to set the needle on five dials placed just within reach. The combined motions of man and room generate a complex pattern of accelerations, including Coriolis accelerations. The dials are set in random order in accordance with taped instructions until either 100 head motions have been made or the endpoint of a mild level of motion sickness (severe malaise) is manifested.

At the time of the first test T1 was experiencing "slight lightheadedness," but this was not increased nor did manifestations of motion sickness appear in completing 100 head motions with the room rotating at 7.5 rpm. The next day T1 was again experiencing lightheadedness. The test was carried out with the room rotating at 10 rpm, and he completed 100 head motions without reaching the endpoint. He did experience dizziness, which was not abolished by closing the eyes, and slight sweating. On the afternoon of the same day the test was repeated with the room rotating at 15 rpm, and the endpoint, severe malaise, was reached after 80 head motions. Symptoms included severe dizziness (lightheadedness was not present prior to rotation), moderate sweating, and moderate nausea; "deep breathing" was observed. Fifteen minutes later the only symptom of consequence remaining was dizziness with eyes open or closed.

The results of the Dial Test indicated that, on one day at least, T1 was susceptible to motion sickness and that the level of susceptibility was somewhat lower than that manifested by normal subjects in the middle third of the normal range. This strongly indicated that T1 had residual function in the vestibular organs. Based on our past experience,

however, we would not have expected symptoms to appear in subjects with similar vestibular residual if the pathologic process was quiescent.

**Earth-horizontal Rotation.** Rotation at constant velocity about an Earth-horizontal axis stimulates the vestibular organs and thus may be used as a provocative test for susceptibility to motion sickness. Tests were conducted on two days. Prior to exposure on the first occasion TI was symptom free. Clockwise (CW) rotation for 90 seconds at 30 rpm did not elicit symptoms, but counterclockwise (CCW) rotation was terminated after 30 seconds because of motion sickness; the manifestations included pallor, sweating, and nausea. On the second occasion prior to exposure TI was experiencing dizziness and a "sensation of intoxication" which was not due to alcohol; exposure to CW and CCW rotation for 90 seconds at 30 rpm did not elicit symptoms of motion sickness.

The fact that TI experienced motion sickness after brief counterclockwise rotation about a horizontal axis on one day and not on another strongly suggests that his susceptibility was related to activity of the pathologic process and not to the fact that "chronic" vestibular disequilibrium was responsible. This inference is further supported by the fact that the nystagmic responses, mentioned above, were normal on the day he did not experience motion sickness but abnormal on the day on which he was highly susceptible. These findings (nystagmus and motion sickness) during horizontal rotation have even more important implications. These are based on the fact that during horizontal rotation, the residual elements in the otolith organs were stimulated in an unusual manner and on the reasonable assumption *vide supra* that the horizontal pair of canals were not functioning. Thus, it would appear that stimulation of the otolith organs evoked motion sickness on the day the pathologic process was active and normal horizontal nystagmus on the day it was quiescent. Inasmuch as the level of the accelerative stimulus was the same, the pathological process either increased TI's susceptibility or contributed to the abnormal pattern of sensory inputs evoking motion sickness.

### III — FOLLOW-UP

Between 1966, when TI left Pensacola, and his discharge from the Army, February 7, 1968, his activities did not involve hard physical work. He did not participate in organized athletics but was active playing with his children, joining "pick-up" games and other sports. He stated that his "depth perception" was not good and felt that it stemmed "at least in part" from his defects. This was noted at twilight and at night, playing football and volley ball, and also while driving. Momentarily, on one occasion when wrestling under water, he lost his awareness of "up" but quickly recovered. Dizzy spells greatly decreased, and at no time did he refrain from any specific activity due to fear of such episodes.

After discharge from the Army, TI returned to his work as a sanitarian, driving a car approximately 1500 miles a month in the course of inspecting health-regulated businesses. TI described his activities and dizzy spells in these words:

I am a junior high school church group sponsor. With this group I have played softball, football, volley ball, swam, ran, and done every other thing you have to do to keep up with the kids. I have mentioned the underwater incident and the depth perception at night problem. Catching fly balls while playing softball is occasionally a problem, especially if I have to move any distance for the catch.

I really cannot remember all of the episodes, in fact, I can remember very few. Actually, only two episodes stand out and that is because of the change in (their) character--- both occurring in 1968, one in the summer and one in the fall. In both instances I was standing. The sensation was that I was tilted to my left at about a 45-degree angle. It was not a sensation of falling or of movement, just suddenly I felt like I was standing sideways on a hill. The episodes were brief and there were no aftereffects.

There was a limited opportunity to conduct some tests in July 1969. Movies were obtained demonstrating differences between TI and a normal subject under challenging conditions, some with eyes open and others with eyes closed. These conditions included a sudden stop after prolonged rotation at velocities up to 50 rpm, making head motions out of the axis of rotation while rotating at 30 rpm, and rail walking. TI was unaffected by procedures that evoked strong abnormal responses in the normal subject; i.e., past-pointing, ataxia, sensations, and motion sickness. On the other hand, TI's performance was far below that of the normal subject on certain items of the ataxia test battery (SR, WOFEO, and rail walking with eyes closed). TI demonstrated excellent ability to catch objects thrown without warning at random points on the perimeter of an imaginary rectangle defined by his extremes of reach (left, right, and overhead) and his shoe tops. He performed well on a bicycle despite not having ridden for 10 years.

The audiogram was normal (Figure 3). Caloric tests, using irrigating temperatures up to 45°C and down to 2.4°C, did not elicit vestibular responses of any kind. Electro-oculograms obtained in connection with the caloric tests revealed neither spontaneous nor positional nystagmus.

Ocular counterrolling was measured twice and the mean values (open circles) are shown in Figure 4. Note that the curve depicting these values is almost identical with the curve based on measurements made on eight normal subjects in the weightless phase of parabolic flight when the otolith organs were (physiologically) deafferented. For comparison, the counterrolling values obtained in a typical test in 1966 are shown.

#### COMMENT

In general, this follow-up period was characterized by: 1) excellent health; 2) a striking decrease in number, then disappearance of dizzy spells; and 3) the virtual loss of all vestibular function. Aside from "the depth perception at night problem,"

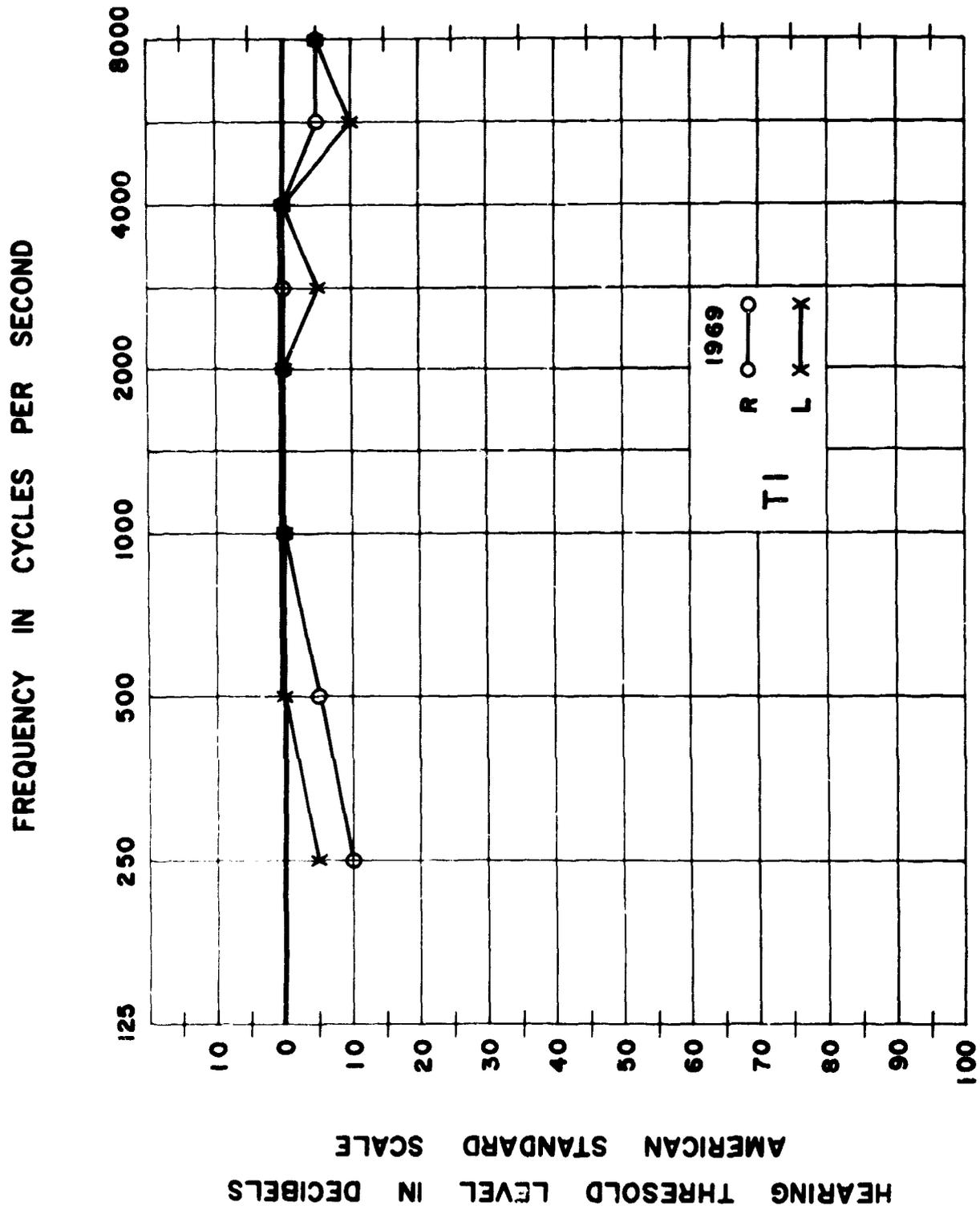


Figure 3  
Hearing threshold level of TI on medical evaluation in 1969.

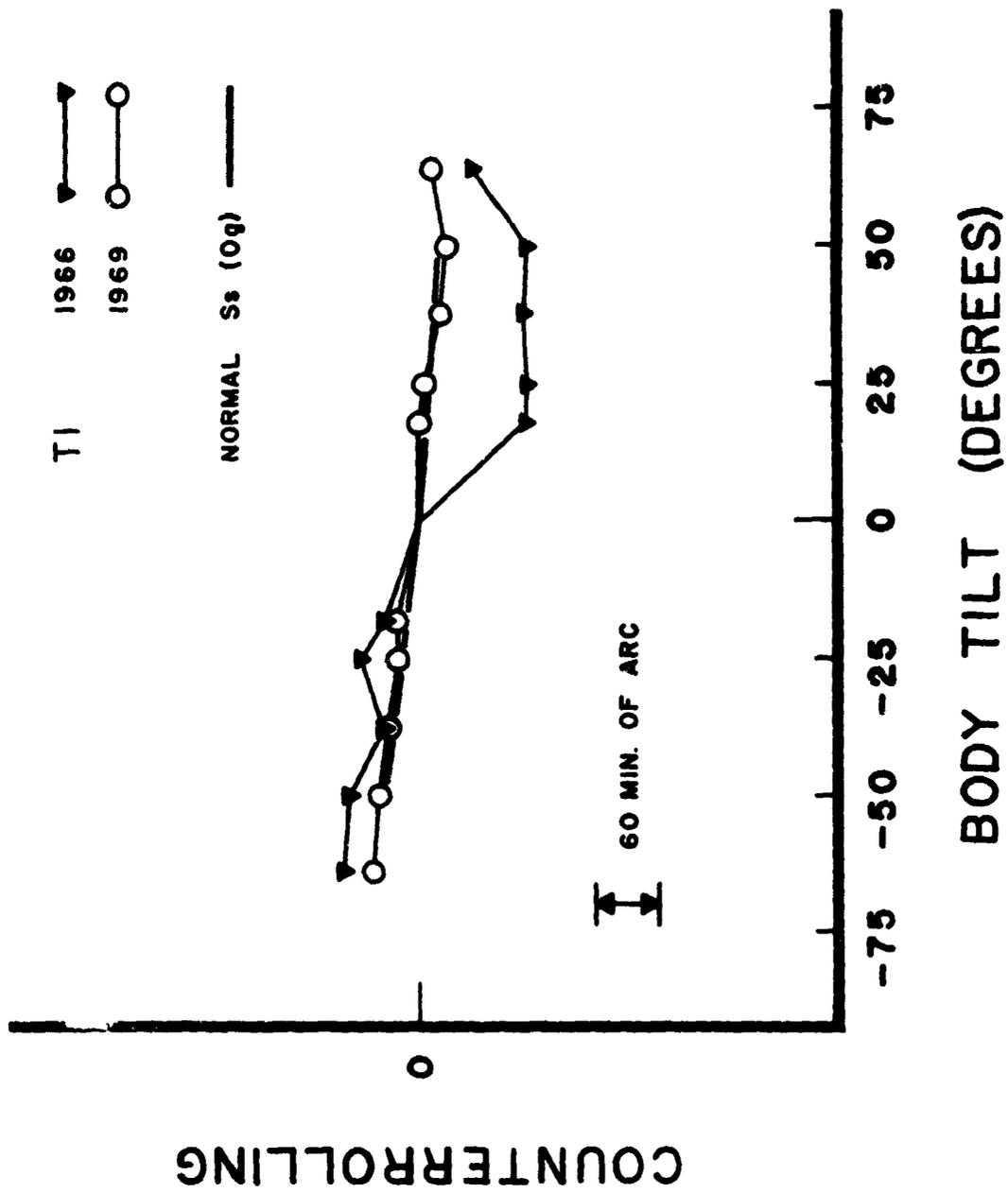


Figure 4  
 Mean ocular counterrolling values (1969) and values obtained in a representative test (1966) on TI compared with the mean values on eight normal subjects in zero g.

TI had no significant complaints although he felt handicapped in the ways described above. On the positive side, he pointed out the advantages of freedom from dizziness, which normal persons may experience in playing certain games, and that freedom from motion sickness is another advantage.

## GENERAL DISCUSSION

It remains briefly to discuss the course of events in this case from three standpoints; namely, diagnosis, vestibular mechanisms, and the role of the vestibular organs in the human economy.

### DIAGNOSIS

Although the onset of TI's affliction is marked by uncertainty, almost surely it was acquired in youth, the pathological process marching slowly but inexorably on a malignant course and terminating in loss of canalicular and otolithic function. The dizzy spells TI experienced between 10 and 15 years of age may have been of vestibular origin, and the repeated attacks of otitis media during this period suggest but do not prove an etiological relationship. There was a change in the character of the attacks from typical vertigo to a geogravic illusion or "leans," which correlated with the order of disappearance first of canal function then otolith function in his late twenties. Despite the extensive bilateral vestibular involvement, there were no manifestations referable to the acoustic labyrinth.

In the pertinent literature we have found only two case reports that in some respects resemble our own. Diamant's patient (etiology unknown) (17) was a shop-hand, 33 years of age, with a recent history of occasional dizzy spells and disequilibrium. His past history was noteworthy only in that 8 years previously he had experienced a severe attack of "giddiness" with prompt recovery. Medical and neurological examinations revealed no definite abnormality. The hearing was normal. Caloric tests (200 ml 8°C) bilaterally resulted in a few "nystagmus-like" deflections. All other vestibular tests, including the galvanic test "with current intensified to the point of pain," pointed to the absence of canal and otolith function. Noteworthy features were the excellent compensation for loss of vestibular function and the brief complaints of vestibular disturbances.

The second patient, reported by Guttich and Stark (18), was a 21-year-old Ensign whose presenting complaint was "difficulty in marching at night," causing him to stagger or fall. Disequilibrium was first noticed during gymnastics toward the end of his elementary schooling. There was also a history of "dizziness" and, under conditions when visual cues were inadequate, disorientation. Otherwise, he had been notably free of disease and disorder, and his family history was noncontributory. The only significant findings in the medical evaluation were referable to the inner ear. The audiogram was normal except for a 60-dB loss at 6000 Hz on the left. There was neither spontaneous nor positional nystagmus; a "significant but small nystagmus of one or two beats" was elicited during the caloric test (12°C for 40 seconds). Additional caloric and rotary tests were confirmatory, but a specific test for otolith function was not reported. Although a diagnosis was made of

"probable congenital absence of the vestibular apparatus," in the light of the findings acquired disease could not be ruled out.

The two patients just described and our own have in common the following: 1) repeated episodes of dizziness relatively early in life, 2) bilateral loss of vestibular function with retention of hearing, 3) freedom from other neurological disturbance or disease, and 4) good general fitness.

TI does not fit into the group of patients designated vestibular neuronitis (19-22) unless the definition is broadened to the point where it has small clinical significance. He cannot be placed in what Coats (22) has designated the "restricted" category of vestibular neuronitis, comprising the only syndrome with clear-cut characteristics; TI would be unique even if placed in the broadly defined category. The absence of cochlear symptoms rules out Meniere's disease and even atypical Meniere's disease in which the diagnostic triad is at first incomplete (23). A suggested descriptive diagnosis in our and similar cases is "idiopathic progressive vestibular degeneration."

#### VESTIBULAR MECHANISMS

At a time when there was good evidence of otolithic function but insufficient evidence to indicate function of the horizontal pair of semicircular canals, TI qualified as a naturally occurring human subject of great value in resolving the question whether the horizontal canals are stimulated by rotation at constant velocity about an Earth-horizontal axis (14,24). On the two occasions when TI was rotated in this manner nystagmus was obtained; in one instance, the response was that anticipated from a normal subject and, in the other instance, somewhat deviant. Almost certainly this nystagmus did not have its genesis in the horizontal canals, implying it was of otolithic origin. The fact that on one occasion TI became motion sick adds to the confirmation. The same explanation probably accounts for the fact that, when some of our L-D subjects with possible residual function of the otoliths but no response to stimulation of the horizontal canals were exposed to Earth-horizontal rotation, nystagmus was elicited in some (14), although not of a normal character or intensity. The fact that Gemandt (25) has demonstrated in cats and monkeys that a mechanical stimulus involving only the utricular macula evokes nystagmus renders untenable the theory that nystagmus can be elicited only by stimulation of the canals.

The findings also emphasize the difficulties and limitations in conducting functional or provocative tests in the presence of overt phenomena indicative of an "irritative" vestibular lesion.

These aspects are pointed up by a comparison between the performance of TI during periods when the pathological process was in an active phase and the responses of our L-D subjects, but particularly those responses of our Ménière's patients (26) with quiescent lesions but far more residual function than TI had. A corollary follows; namely, that residual insufficient for purposes of contributing to behavioral responses may, when stimulated by pathologic processes, evoke reflex vestibular perturbations and motion sickness.

It has been mentioned above that TI experienced relatively few attacks of severe vertigo, considering the great losses of vestibular function. It is suggested that the severity of symptoms, and especially those characteristic of motion sickness, reflects, in part at least, the basic or innate susceptibility of the person afflicted. TI was highly insusceptible to motion sickness at times when it could be presumed that the disease process either had not yet appeared or was quiescent. Individual differences in susceptibility to motion sickness (history) might offer a useful clue in evaluating the significance of symptoms of patients with vestibular disease; e.g., in the case of two patients with vestibular involvement of similar type and severity, one with low susceptibility might complain of dizziness and the other of dizziness, nausea, and vomiting.

#### ROLE OF VESTIBULAR FUNCTIONS IN HUMAN ECONOMY

The fact that prior to the fortuitous detection of labyrinthine disease, TI never made any complaints referable to a deficit in vestibular function *per se* and the fact that he met and passed all of the test conditions in real life *incidental* to acquiring a college education, earning a living, and enjoying participation in a wide variety of sports, have important implications. The most important inference is that the vestibular organs are not essential to what might be termed good health and general fitness (including muscular fitness) for most of life's activities. Although TI's scores in the postural tests revealed the characteristically sharp limitations of persons who must rely largely on visual cues and somatic sensory systems, he compensated for these deficits to the point where neither he nor others detected any deficiency. The fact that he did not stumble, fall, or otherwise perform in a clumsy manner implied that he learned to operate with something in reserve to meet unexpected demands on his skills. Only one specific way, presumably among many (27) in which compensation could be effected, was demonstrated. TI, in common with most subjects with labyrinthine defects, manifests (with eyes open) little or no improvement in test scores with practice, whereas many normal subjects do (8), implying that TI exploited visual cues almost to the fullest extent possible. It should be pointed out that TI probably was not representative of a random group and that his innate talent and early acquisition of skills in sports helped in compensating for losses of vestibular function. The findings in this case do point out what might be possible in similar instances and emphasize that, in man, the unique role of the vestibular organs (for which there is no compensation) is relatively small. If a large general population of young people were screened or if all complaints of dizziness in such a group were carefully evaluated, more cases like ours would be uncovered.

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13. ABSTRACT

**This report presents the case of a young man, T1, who experienced episodes of dizziness and demonstrated the rare combination of bilateral loss of nonacoustic function with retention of normal hearing. Persistent dizzy spells first appeared in childhood and were usually mild, but, on rare occasions, he was handicapped by brief severe attacks of vertigo. T1 was nearly 26 years of age before these episodes were shown to be of vestibular origin; yet, during the preceding years he never complained of symptoms attributable to loss of function, as distinct from perturbed vestibular function. At that time residual function was substantial in the otolith organs but minimal and nil in the vertical and horizontal canals, respectively. When last seen at age 29, even otolith function was lost; this was associated with the gradual disappearance of dizzy spells. The diagnosis, idiopathic progressive vestibular degeneration, was made by exclusion. The notable conclusion to be drawn from this patient was not the fact that he had dizzy spells of vestibular origin, but the fact that loss of vestibular function per se was not incompatible with leading a life regarded by himself, his family, and friends as normal.**

10. KEY WORDS	LINK A		LINK B		LINK C	
	ROLE	WT	ROLE	WT	ROLE	WT
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Vestibular apparatus						
Vestibular function tests						
Hearing						
Idiopathic vestibular degeneration						