MESENCEPHALITIS AND RHOMBENCEPHALITIS

BY

E. R. BICKERSTAFF, M.D., M.R.C.P.

Senior Registrar, Department of Neurology, United Birmingham Hospitals

AND

P. C. P. CLOAKE, B.Sc., M.D., F.R.C.P., D.P.H. D.P.M.

Professor of Neurology, University of Birmingham

Despite the advances of the recent decades in scientific methods of investigation, conditions are not infrequently seen, particularly in neurological practice, which fall outside the well-recognized syndromes and in which it is possible to hazard only a guess at the aetiological and pathological processes involved. If the patient recovers the exact explanation may never be certain, but one experience of such a condition may enable it to be recognized again, and, even in the face of ignorance about its nature, some idea of its likely course and prognosis may be formed.

This was at one time the only method available to physicians, and it is not surprising that occasionally quite different morbid processes were classified under the same clinical heading.

It is possible that in classing together the three cases recorded below, all of which were seen in this unit within eighteen months, one may be guilty of the same error, but their clinical features and course were so remarkably similar that it seemed highly probable that the same or a closely related pathological process was involved. The picture they presented was striking, and its exact form does not seem to have been previously described.

The purpose of this paper is to draw attention to this type of case, to the possibility of complete recovery from an apparently moribund condition with total bulbar palsy, and to the necessity for sparing no effort to combat secondary infection and to maintain nutrition during this period.

If it is correct to regard these cases as examples of the same condition, two showed it in its most severe form, while the third was milder and was arrested at an intermediate stage.

Case 1

A married warehouse forewoman aged 24 was admitted to hospital on January 15, 1949. Her previous history and family history were not relevant. She had had malaise with vague muscular aching for two weeks, and for one week increasing drowsiness with drooping of the lids, more marked on the left. The left corner of her mouth had dropped, and her gait had become a little unsteady. Four days later she saw double on lateral gaze, her speech became slurred, she had slight difficulty in swallowing, and felt numbness of the left face, arm, and leg. The drowsiness then became overpowering; there was difficulty in moving the eyes, and impairment of hearing.

General examination on admission revealed no abnormality, though she had a pyrexia of 99–99.6° F. (37.2–37.55° C.), and pulse rate of 100–120. There was no aural disease. She was very drowsy, but was easily roused to full co-operation, lapsing into sleep again as soon as conversation ceased. The fundi and fields were normal. There was marked bilateral ptosis, and gross defect of conjugate ocular movement upwards, laterally, and to a lesser degree on convergence, downward movement being normal. Pupillary reactions were present. Fine and fast nystagmus to the right was present, slower and coarser to the left. The left masseter and temporalis were weak, and sensation was impaired over the left face. There was a bilateral facial weakness. Examination of the remainder of the nervous system showed left-sided ataxia, weakness of the left abdominal reflexes, an equivocal left plantar reflex, and left-sided hypalgesia.

Investigations.—*X*-ray films of skull and chest were normal. Urine was normal. Cerebrospinal fluid: pressure, 125 mm.; normal hydrodynamics and biochemistry; 26 cells per c.mm., 90% mononuclear. White blood count 16,000, 84% polymorphonuclears. Electroencephalogram: bursts of high-voltage delta activity in all right-sided leads with phase-reversal in the right temporal region.

From the time of admission for four weeks there was a slow, steady deterioration, the ptosis becoming complete and the eyes fixed except for some downward movement. Bilateral total paralysis of jaw, facial, palatal, and pharyngeal muscles resulted in inability to hold her mouth closed or to lift her head off the pillow. She was unable to swallow or to speak, and deafness was marked. The remainder of the body appeared unaffected and her mental state was quite clear, though to a casual glance she appeared moribund. Nasopharyngeal secretions had constantly to be sucked out, but she would often attend to this herself. She was fed by oesophageal tube twice daily, and the constantly threatening inhalational bronchopneumonia was combated by full chemotherapy and penicillin.

After remaining in this state for three weeks, with numerous episodes of respiratory obstruction, each thought to be her last, she made an indistinguishable sound, and three days later to our astonishment could enunciate recognizable words, while some movement returned to the eyes. After a week she was able to speak sentences, in 10 days was able to swallow fluids, and solids four days later; eye movements were limited only on upward gaze.

There was a slow and uninterrupted improvement, but as she became more mobile it was apparent that, though eye movements were normal, and all the motor weakness had disappeared, she had developed a Parkinsonian facies with tremor of the head, and mild cogwheel rigidity of the limbs with tremor. The cerebrospinal fluid on March 31 was normal apart from the Lange curve, which read 4322100000.

Her further progress was followed in the out-patient department, and the Parkinsonian features gradually disappeared. By February, 1950, there was slight immobility only of the face, and six months later she was in all respects quite normal.

Comment.—On admission the sensory, cerebellar, and mild pyramidal signs were all on the same side, and, with the gross defects of conjugate ocular movements, pointed to the main lesion being in the tegmentum of the midbrain, with evidence of involvement of lower centres developing later. It was striking how, at the height of the illness, when the patient was unable to see, speak, swallow, or expectorate the secretions which could be heard rattling in her throat, her mental state was perfectly clear and the power in her limbs excellent. She was very eager to listen to the wireless, and her deafness necessitated its being extremely loud. It was an unforgettable sight to see an apparently moribund patient, her wireless set at full volume, occasionally turning on the motor of the sucker to clear her own nasopharynx.

Case 2

A male 36-year-old works inspector had a history of general malaise and recurrent frontal headache for seven days, and increasing drowsiness for two days during which both lids tended to droop. On March 21,

1950, he tried to play table-tennis, but was too sleepy, a little unsteady, and unable to follow the ball quickly with his eyes. On the way home he saw double on lateral gaze. In the next two days all symptoms grew worse and he attended an eye hospital, where he was noted to have bilateral ptosis, non-reacting semi-dilated pupils, and slight weakness of lateral eye movements. The drowsiness and ptosis grew worse and he was admitted to the Queen Elizabeth Hospital on March 27, where it was found that on being roused he was perfectly sensible and co-operative, but tended to sleep again if his attention was not held. Marked bilateral ptosis was present; the pupils were semi-dilated and fixed; there was total palsy of conjugate movement upwards and of convergence, lateral conjugate movement was grossly defective, but downward movement was normal. Nystagmus to the left was present. There was a slight bilateral ataxia of the limbs and all tendon reflexes were abolished. Other systems were normal. His blood pressure was 150/85.

Investigations.—Blood count and W.R. normal. *X*-ray films of skull and chest normal. Cerebrospinal fluid: pressure 125 mm.; hydrodynamics normal; containing less than one cell per c.mm.; protein, 40 mg. per 100 ml.

In the next week there was little change, the drowsiness continuing, but on April 7 he was seen to have a total right facial paralysis. In three days this was better and a steady improvement ensued, the ataxia disappearing first, the pupillary reactions next returning, and convergence and lateral deviation approaching normal. The cerebrospinal fluid on April 11 showed 77 mg. of protein per 100 ml. and a normal cell count. Lange 2222111000.

A month after admission the cranial nerves were normal, except for limitation of ocular movements upwards, and the tendon reflexes were just perceptible. Four weeks later the reflexes were normal, and after a further two months the eye movements became entirely normal.

Comment.—It appeared at first that the march of events was following a course similar to that in Case 1, and a period of grave illness was expected. Progression stopped, however, when facial paralysis had occurred, and recovery took place over a period of months. A striking feature was the abolition of tendon reflexes, which recovered *pari passu* with the cranial nerves.

Case 3

A 24-year-old married woman, with two children, was admitted to hospital on June 1, 1950, for removal of an ovarian cyst.

On admission, apart from symptoms relevant to the ovarian cyst, she had had a throbbing frontal headache for one week. On June 6 she was missed from her ward, and no trace of her could be found. A lavatory door was seen to be locked; no response being obtained from within, it was forced open and she was found lying on the floor, apparently recovering consciousness after a faint. She had been vomiting, but not incontinent, and as there were no abnormal signs the oophorectomy was performed four days later. The cyst was simple. After full recovery from anaesthesia she remained drowsy, and ran a slight temperature. On June 11 the drowsiness was worse, her speech became slurred, and she saw double to the right. During the next week the drowsiness increased, and on June 19 there was obvious dysarthria. A secondary wound suture was performed on the 20th. Two days later she had a general convulsion, following which marked drowsiness, dysarthria, diplopia in all directions, and a doubtful bilateral facial weakness occurred. Full blood count, Wassermann test, and skull and chest radiographs were normal. Cerebrospinal fluid pressure was 75 mm., and the fluid contained 153 mononuclear cells per c.mm. with a normal biochemistry.

Three days later a second convulsion occurred, and the day after this the electroencephalogram showed marked widespread abnormality with delta and theta activity. The cerebrospinal fluid now contained 67 mononuclear cells per c.mm. and the Lange curve was 2221100000.

There was now a steady deterioration, the drowsiness increasing and the dysarthria progressing to anarthria. Bilateral ptosis developed and conjugate ocular deviation upwards and laterally became defective, while convergence was impossible. Nystagmus to right and left was present. Bilateral facial, palatal, and pharyngeal palsy then developed, the motor element of the fifth nerve became paralysed, and the patient was unable to support her jaw and later to raise her head. Her eyes then became fixed and some deafness developed. Slight ataxia was the only abnormality in the rest of the nervous system.

For a week she remained in this state, being fed by oesophageal tube, needing constant suction to clear the nasopharyngeal secretions, and appearing moribund; but there was then a gradual return of motor function, first in the movements of the eyes, then in the face and jaw, and, finally, in swallowing. It became apparent, however, that a gross psychotic state was developing, with alternating periods of maniacal excitement and katatonia. By August there was return of normal power, but speech was difficult to understand, partly owing to the animal-like behaviour and noises which were features of her mental abnormality. After transferring her to a mental hospital her condition remained unchanged for several weeks. A slow but steady improvement then took place, and by the end of four months, apart from a slight dysarthria, she was mentally and physically perfectly normal.

Comment.—Owing to her admission to hospital for other reasons it was possible to watch the development of this patient's illness from the onset, and it was remarkably similar to the other two cases, except that she had in addition two convulsions and possibly a third. From previous experience the course of her disease was foreseen, and her appearance and neurological state when at its worst were identical with those of Case 1.

Clinical Features

Onset and Development.—The gradual progression with evidence of daily more widespread involvement of the brain stem was one of the characteristic features. In Case 1 there was slow progression for four weeks, in Case 2 for two weeks, and in Case 3 for four weeks, until in Cases 1 and 3 there was almost total suppression of function of the third to the twelfth cranial nerves. A stationary period of three weeks, one week, and one week respectively was then followed by spontaneous remission over several months until complete functional recovery was achieved.

General Symptoms.—Each patient had been unwell for a few weeks prior to the onset. One had pain in the back, another in the abdomen (attributed to the presence of an ovarian cyst). All three showed early mild pyrexia —99–99.8° F. (37.2–37.7° C.)—and two suffered from frontal headache for a week before the onset of other symptoms. There was no very severe headache, no vomiting, delirium, signs of meningeal irritation, or photophobia, nor was there any history of prodromal infections, skin eruptions, or recent immunization procedures.

Drowsiness.—This was a prominent early symptom in all three cases. It was that type of drowsiness seen in lesions of the midbrain, when the patient is easily rousable, intelligent and co-operative when roused, but rapidly sinks again into sleep when attention is no longer held.

Oculomotor Disturbances.—Ptosis was marked from an early stage in two patients and developed after a

week in the third. Combined with the drowsiness and facial paralyses, it gave the striking similarity to the appearance of the three patients. Diplopia then occurred, mainly on lateral gaze, and was followed by difficulty in moving the eyes, so that the whole head was turned to follow moving objects; it was objectively demonstrable as a gradually increasing failure of conjugate ocular deviation. Upward movement and convergence were most affected, lateral movement for a shorter time and to lesser degree, and downward movement hardly at all. In each case the eyes were practically fixed for a variable period, and recovery occurred in reverse order of development.

Lower Brain-stem Involvement.—In two patients total bilateral paralysis of the motor part of the fifth nerve occurred, it was impossible for them to support their jaws, and in one sensation on one side of the face was defective. Bilateral facial palsy developed in two and right-sided palsy in one. In two cases the muscles of the palate, pharynx, tongue, and sternomastoids then became so weak that the patients had to be fed by oesophageal tube, and were unable to raise their heads from the pillow. Dysarthria accompanied this, and proceeded to a total anarthria. Bilateral deafness, developing at the same time, made contact with the patients increasingly difficult. In one case this phase lasted for three weeks, but in the second for one week only, and at no time was there evidence of involvement of respiratory or circulatory centres.

Remainder of the Nervous System.—This was comparatively little affected. No motor weakness of trunk or limbs was seen at any time, and signs of pyramidal-tract involvement were seen only in one case, and then very slight and doubtful. All three showed a mild but recognizable ataxia of arms and legs, and in one patient there was total abolition of all tendon reflexes. Case 1 had a hemihypalgesia affecting the whole body on the left side.

Evidence of Cortical Involvement.—Two, possibly three, major convulsions occurred in one patient, and in two the electroencephalogram was abnormal, the abnormality disappearing in one as the patient recovered. The late development of a psychotic state in the third was not marked by definite symptoms recognizable as cortical in origin, but may none the less have been due to disturbance of cortical or diencephalic function, or simply to a general disorder of mind resulting from grave and terrifying illness.

Laboratory Investigations. — The electroencephalographic findings have been mentioned. All radiographic studies were normal. The pressure of the cerebrospinal fluid was normal, but in two cases there was a marked mononuclear pleocytosis without biochemical change, while in the third the protein rose without cellular reaction. A paretic Lange reaction was seen in all three.

Course and Sequelae.—Slow neurological recovery over a varying period was seen in each case, the time taken for complete restitution of function varying from two to ten months. In Case 1 there was a period of typical Parkinsonism lasting two months, which also completely recovered. In Case 2 there were no sequelae, and though in Case 3 neurological recovery was complete the development of a psychotic state necessitated institutional treatment. This, however, was only temporary, and mental recovery was also perfect.

Nomenclature

A purely descriptive term has been employed to classify this condition. It is unfortunate that an "encephalitis" is so often loosely diagnosed when a grave but unexplained cerebral disturbance is seen, and, despite the fact that the pathological lesions in the most famous of all encephalitides—the encephalitis lethargica of von Economo—were particularly to be found in the substantia nigra and periaqueductal region of the midbrain (von Economo, 1931; Weil, 1946; Scheinker, 1947; Lichtenstein, 1949), it is often assumed that the term signifies mainly involvement of the hemispheres, and bulbar poliomyelitis and polio-encephalitis are not necessarily considered synonymous. Though the hemispheres appear to have been affected in at least two of these cases, the main brunt of the disorder was borne by the midbrain at first and later the lower brain stem. The terms "mesencephalitis" and "rhombencephalitis" have therefore been suggested.

The suffix "itis" may be open to criticism. The alternative would be "mesencephalopathy"—a term which is neither euphonious nor indicative of the belief that we are dealing with an inflammatory process.

Discussion

It is open to question whether anything is to be gained by describing a clinical picture entirely without pathological material, and in the face of the ever-present possibility that one is dealing with three entirely different conditions. The similarity of these cases at one stage of their disease was, however, most striking. The bilateral ptosis with fixed eyeballs and bilateral facial palsy and the gross bulbar palsy in Cases 1 and 3 gave to these so moribund an appearance that recovery seemed impossible, yet it could be seen that excellent power remained in the limbs, and if they were able to make their wishes known sphincteric control was perfect.

It may be said that this similarity was due only to the site of the lesion in each case and that the pathological processes may have been entirely different. It seemed, however, to be more than this, for, apart from the anatomical structures involved, the course of the disease was so similar, with its division into three stages characterized by gradual development involving first the midbrain and then the lower brain stem; a variable period of maximal disability in which two of the patients appeared moribund; and then even more gradual regression to final total neurological recovery. The experience of the first case enabled us, when seeing the other two, to give a prognosis and adopt a course of nursing care based on the assumption that there would be a period of grave disability which, if all measures were taken to maintain nutrition at its highest level even when the patients' condition appeared hopeless, would be followed by gradual complete recovery. In one case the progression seemed to be arrested at an intermediate stage, but in the other the train of events we anticipated happened, and it was disappointing, when complete neurological recovery had occurred, that the development of a psychotic state necessitated treatment in a mental institution, even this, however, totally recovered.

The aetiology is quite unknown. The gradual development to maximal disability with subsequent recovery seems to eliminate vascular thromboses, haemorrhages, or emboli. The total recovery argues against neoplasia, and this makes equally unlikely a pyogenic abscess,

particularly in the absence of any infective process elsewhere. "Mesencephalitis syphilitica" (Wilson and Cobb, 1924) appears to be excluded by the repeatedly negative blood and cerebrospinal fluid Wassermann reactions, and there was no exposure to known neurotoxic agents.

The "acute superior haemorrhagic polio-encephalitis" of Wernicke is associated with dietary deficiencies—either primary or secondary—has a more acute onset, and, though dysarthria and seventh-nerve palsy may occur (Spillane, 1947), the gross bulbar palsies seen in Cases 1 and 3 and the cerebrospinal fluid changes do not occur.

A very similar picture may be caused by botulism, but the onset is again more acute and the spinal fluid normal (Kinnier Wilson, 1940). There was no history of exposure to infection or of gastro-intestinal disturbance.

Hunter (1949) described an encephalopathy occurring after anaesthesia, which might be considered in Case 3; but that patient's first manifestations appeared before the operation, the cases described were all fatal, and none showed any localization in the brain stem, the clinical picture bearing no resemblance to these cases.

Practically all the common infectious diseases which have an encephalitis as a complication have been known to show clinical involvement of the brain stem (Kinnier Wilson, 1940), but the gross bulbar palsies are not seen, and there was no history of affection with or exposure to any of these conditions.

The two most attractive possibilities remaining are the demyelinating conditions, such as disseminated sclerosis and the primary infections of the nervous system presumed to be of virus origin. The course of the disease, the defects of ocular movement, the ataxia, and the spinal fluid changes could be explained by a plaque of sclerotic tissue developing in the tectum of the midbrain, but the absence of pyramidal-tract involvement and the predominance in the second stage of lower motor neurone paralyses make this diagnosis very unlikely.

All the various features, however, could be explained by an infective process of virus origin, the main brunt of which was borne in the early stages by the midbrain, but which later extended to involve the lower brain stem, reached its maximum, and then recovered. In Case 2 there was in addition abolition of the tendon reflexes, which totally recovered; and, indeed, the gradual onset, the period of maximal disability, and the subsequent gradual recovery are very similar to the course of the so-called infective polyneuritides which are widely accepted, though without proof, as being due to virus infection. In this relation it will be noted that a mild albumino-cytologic dissociation was present in the cerebrospinal fluid in Case 2.

It will be recalled that Guillain and Kreis (1937) described a "mesencephalic type of the Guillain–Barré syndrome"—but this was so-called because the clinical evidence suggested that certain of the cranial nerves originating in the brain stem were involved, and there was not the involvement of the parenchyma of the brain stem as suggested in these cases, nor was there a bulbar palsy.

Several unusual forms of encephalitis have been seen in this country in recent years, and four main groups, classified by pathological examination, have been reviewed by Greenfield (1950). Predominant involvement of the brain stem was not a feature, and they ended fatally.

In the 1941 North Dakota epidemic of encephalitis there was pathological evidence of severe brain-stem involvement, but the onset was very sudden, with high fever and neck rigidity, and the whole clinical picture was quite different from our cases (Weil and Breslich, 1942), remarks which apply also to the 1938 epidemic in the same area (Breslich *et al.*, 1939).

Pathologically the mesencephalon was most severely affected in the St. Louis epidemic of 1933, but here again the onset was marked by signs of meningeal irritation, and oculomotor palsies were absent throughout (Weil, 1934).

Might our cases, however, be examples of encephalitis lethargica? All three presented a phase similar to that seen frequently during the major epidemics, and it will be recalled that Case 1, during the phase of recovery, passed through a period of typical Parkinsonism. Von Economo (1931) stresses the great variety of clinical manifestations which may be produced by the disease which bears his name, and writes: "Illnesses of the nature of acute bulbar paralysis, which terminate fatally or end in recovery, have not only been observed in combination with the three 'basic' forms but also as isolated occurrence. These disturbances were usually transitory and benign." He refers also to epidemics of bulbar paralysis in Germany—but in these cases the paralysis was only partial, and it is not clear on what grounds many of the cases were distinguished from poliomyelitis. Psychotic phenomena, such as were seen in Case 3, were also common during the major epidemics.

In our cases, however, even the Parkinsonism in Case 1 completely cleared up, the onset was always gradual, with little or no systemic disturbance, and no reversal of sleep or respiratory rhythm occurred. There have been no proved cases of the disease in this locality, and bulbar symptoms of such severity, which even if recorded are very rare (Brain, 1947), were predominant in Cases 1 and 3.

The severity of the bulbar signs, and the predominance of the lower motor neurone involvement, are very similar to that seen in bulbar poliomyelitis. The gradual onset and progression, the absence of meningitic signs in the earliest stages, the great prominence of the supranuclear or nuclear ocular disturbances, the absence of respiratory and circulatory centre involvement in the presence of gross medullary disease, the paucity of the cerebrospinal fluid changes, and the total recovery of the paralysed muscles without wasting are very unlike the ordinary picture of poliomyelitis.

It is interesting, however, that the clinical picture described above was seen for the first time in this area during the years when, also for the first time, poliomyelitis has become a serious problem. Outbreaks of presumed virus disorders of the nervous system during or in relation to outbreaks of poliomyelitis have been observed before (Ottonello, 1939; De Sanctis and Green, 1942; Stapleton, 1949). One occasionally hears of patients who develop some strange apparently infective neurological disorder after contact with poliomyelitis (Sandifer, 1950). The virus responsible for this disease is now known to be divisible into at least three main groups with different immunological patterns (Bodian *et al.*, 1949), and infection with one group does not give

immunity to reinfection with another (Bodian, 1949). The full extent of these groups is not yet known, and it seems not impossible that infection with a virus of attenuated virulence might produce a condition similar in many ways to poliomyelitis but differing in certain features, and particularly in course. Alternatively, is it possible that previous mild subclinical infection might greatly modify the course in a manner similar to the modification of smallpox produced by previous vaccination? Problems such as this will remain unsolved until methods of research into virus diseases are simpler and more generally available; but there exists a wide field for investigation in this respect into many of the obscure transient disorders of the nervous system, in which the diagnosis is so often, as in these cases, speculative.

Summary

Three cases are described of a condition characterized by drowsiness, total oculomotor palsy, facial palsy, ataxia, and, in two, total bulbar paralysis and anarthria.

All the cases progressed slowly to maximal disability, two of the patients becoming moribund. They remained at this stage for variable periods, and slowly improved until total neurological recovery was attained.

The differential diagnosis is discussed and a possible relationship to poliomyelitis considered.

We wish to express our thanks to Mr. P. Jameson Evans for referring Case 2.

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The Executive Board of the World Health Organization ended its eighth session on June 8 and decided to convene the next session in Geneva, beginning January 21, 1952. The fifth World Health Assembly, commencing May 5, 1952, in Geneva, will hold technical discussions on the economic value of preventive medicine and on the methods of health protection for local areas, as determined by the health, social, and economic needs of the population. The level of expenditure for 1951 was raised to $6,497,401 to provide for relief to the civilian population in Korea and to Turkish refugees from Bulgaria. Emergency action was taken to help prevent epidemics in the famine-stricken areas of India; a sum of $30,000 was appropriated to purchase medical supplies for the prevention of cholera and malaria outbreaks in North Bihar. British Somaliland was excluded from the African yellow fever area. The Executive Board requested preliminary work on four new international health regulations, to deal with the control of malaria insect vectors in international traffic, sanitation standards for airports, the international pharmacopoeia, and legal protection of international non-proprietary names for drugs.

AORTIC ANEURYSM ASSOCIATED WITH ARACHNODACTYLY

BY

M. F. MOSES, M.B., B.S., D.C.H.
Late Postgraduate Registrar, Central Middlesex Hospital

Marfan (1896) published the first report of a congenital deformity of the limbs, especially of the distal parts, with lengthening and narrowing of the bones. The name arachnodactyly (spider fingers) was suggested by Achard (1902). In addition to the various skeletal deformities, anomalies of the eye, external ear, and cardiovascular system have been described. Cardiovascular abnormality was noted by Rados (1942) in 64 out of 204 cases and by Ross (1949) in just over a fifth of 117 cases. The number of reported necropsies is small. Tobin *et al.* (1947) reviewed 12 cases and added two of their own. Reynolds (1950) had found seven more reports and added two of his own. There were also single case reports by Strayhorn and Wells (1947), Spickard (1948), and Vivas-Salas and Sansón (1948).

In half of the necropsies typical aortic lesions were present. Baer *et al.* (1943) and Tobin *et al.* (1947) reported two cases each. Weill (1932), Etter and Glover (1943), Vejdovsky (1946), Uyeyama *et al.* (1947), Strayhorn and Wells (1947), Spickard (1948), Lutman and Neel (1949), Lindeboom and Bouwer (1949), and Reynolds (1950), each reported a single case. In nine of these 13 cases the histology of the aorta was reported.

Two more cases of arachnodactyly with associated aortic lesions are reported here, together with post-mortem and histology findings.

Case 1

The patient, a 52-year-old unmarried woman, was admitted to hospital on September 10, 1949, on account of dyspnoea. Her mother and father were not consanguineous. They died aged about 70 years. The six other children were normal. In her infancy bilateral dislocation of the lens was noted, and in early childhood several operations on her eyes were performed, since when she had worn glasses without disability. She gave no history of rheumatic fever or chorea. She had felt well and worked as a housekeeper until February, 1948, since when she had experienced paroxysms of nocturnal dyspnoea. Dyspnoea on exertion became severe. She also complained of occasional retrosternal pain, palpitation, increasing lassitude, loss of appetite, loss of weight, and recent nocturia.

On examination she appeared a tall, thin woman with a pained expression and a malar flush. Her limbs, hands, and feet were long and slender, with bilateral contractures of the toes. There was a slight dorsal kyphoscoliosis. Her teeth were irregularly spaced, and the palate was high and arched. Her ears were large and protruding.

Measurements were: weight, 8 st. 5 lb. (53.1 kg.); height, 5 ft. 6 in. (167.6 cm.); span, 5 ft. 9 in. (175.3 cm.)—normally the span is less than the height; head length, 17.5 cm.; head breadth, 15.4 cm., making the cephalix index 0.88. Thus there was a degree of brachycephaly, in which the head is short and broad with the skull breadth at least four-fifths of the length. In most of the recorded cases the skull is dolichocephalic, or long-headed. There was scanty subcutaneous tissue, and hypotonic musculature. Both eyes showed shimmering of the iris after sudden movement of the head (iridodonesis). This is secondary to subluxation of the lens, leaving the iris unsupported. Visual acuity was right eye 6/60, left eye 6/9 with glasses.