and once in a left haemothorax. In two cases an associated patent foramen ovale was reported by Baer *et al.* (1943).

In the 13 cases without aortic defects, three had normal hearts, five had patent foramen ovale, one had a patent ductus, three had mitral valve deformity, one of which died from subacute bacterial endocarditis, and one had aortic incompetence which was probably rheumatic. Of these cases six patients died under 28 months of age, whereas all those with aortic lesions lived beyond the first decade.

It has been assumed that the cardiovascular lesion associated with arachnoidactyly is most commonly a patent foramen ovale. However, analysis of the 26 reported necropsies together with the two cases presented here shows that aortic defects were present in over half of the total and in over two-thirds of the adult cases.

Arachnoidactyly often presents with subluxation of the lens, and Lloyd (1948) suggested that the condition be assumed to be present until shown otherwise. Tobin *et al.* (1947) advised that the formes frustes be considered whenever a dissecting aneurysm or unexplained aortic dilatation is encountered.

The histology of the aorta always showed deficiency and rupture of the elastic fibres in the media, which usually contained cystic spaces. Tobin *et al.* (1947) likened the appearance to medionecrosis arotae idiopathica cystica, described by Erdheim (1930). Baer *et al.* (1943) concluded that the condition represented a congenital maldevelopment. Uyeyama *et al.* (1947) suggested a primary anomaly of the vasa vasorum, which is supported by the experiments of Schlichter *et al.* (1949).

Summary

Two cases of arachnoidactyly with aneurysmal dilatation of the ascending aorta and multiple aortic tears, without dissecting aneurysm or rupture, are reported.

In cases of arachnoidactyly with cardiovascular abnormality it is suggested that the aortic defects occur as often as the better-known congenital heart lesions, such as patent foramen ovale. In the adult, aortic incompetence due to dilatation of the aortic ring and ascending aorta constitutes the usually associated cardiovascular anomaly.

These aortic lesions, though resembling the results of acquired disease, are probably following upon a progressive weakening in an inherited congenital malformation of the aortic media.

I wish to thank Dr. H. Joules for his interest and permission to report Case 1; Professor L. S. Penrose for his review of this case; and Dr. R. Asher for permission to report Case 2. I am most grateful to Dr. Walter Pagel and Dr. Norman Ashton for their post-mortem and histological reports; to Dr. Samuel Oram and Dr. Keith Ball for their helpful advice; and to Miss Beryl Burnett for the clinical photographs and the photomicrographs.

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PERSISTENT ANXIETY AND TACHYCARDIA SUCCESSFULLY TREATED BY PREFRONTAL LEUCOTOMY

BY

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The patient whose case is described below suffered from severe and continued anxiety, with constant awareness of persistent tachycardia, for 18 months before operation. Throughout this period her case was personally managed by me. In spite of energetic psychological and physical treatment the illness increased in intensity, and for the last six months she had become bedridden. Prefrontal leucotomy produced a dramatic improvement. She was discharged seven weeks after operation, and five weeks later was doing all her domestic duties adequately. One year after the operation she has remained well. Her mental equilibrium has been restored at the price of the familiar personality changes associated with leucotomy. These have not seriously impaired her functional efficiency. Both the patient and her husband believe the operation well justified.

Case Report

The patient was a married woman aged 35. Eight weeks before admission (18 months before operation) she suddenly became aware of the rapid beating of her heart. She was panic-stricken by the thought that she had heart disease and was going to die. Her doctor found no organic disease and told her that her heart was sound, but she could not accept this or the reassurance of a specialist who found no abnormality apart from tachycardia. Her anxiety increased and she was admitted to hospital.

Her weight was 7 st. 1½ lb. (45 kg.) and her height 5 ft. 1½ in. (156 cm.). She was lean, but denied loss of weight, and there was no objective evidence of it. The pulse rate at rest varied between 110 and 120, rising to 150 on slight exertion. The blood pressure was 130/85. The heart was normal. There was no exophthalmos or thyroid enlargement. Examination of the chest was negative. Clinical examination revealed no abnormalities in other systems.

Special Investigations.—The basal metabolic rate, estimated on five occasions, gave the following readings: +58, +35, +40, +56, +50. Electrocardiograms were repeated on several occasions, with negative results. Thyroid function assessed by tracer methods with radioactive iodine gave normal readings. All other routine and relevant special investigations were negative: these included examination of the urine, blood, blood sedimentation rate, blood cholesterol, blood Wassermann and Kahn, radiograph of the chest and heart, and radiological investigation to exclude retrosternal goitre.

Family History.—The mother was described as an over-anxious type in impaired health who often complained of her heart. The father was a semi-invalid, following a head injury. He suffered from head pains, depression, and

irritability. The patient was sixth in a family of eight. In 1941 a sister aged 33 died suddenly after a cerebrovascular accident while the patient was living with her. In 1943 a sister aged 35 died suddenly of cerebral embolism, 11 days after childbirth. The patient adopted this sister's child, who was subsequently found to have congenital heart disease. A third sister had had many operations and several miscarriages, her general health was impaired, and she was unable to do much. Two remaining sisters were fairly fit, though both had suffered from "heart trouble" and one had had her activities restricted from childhood on this account. Two brothers were healthy.

Previous History.—Her childhood and school days were normal. Nervous traits were denied. She was of average intelligence and mixed well. Her work record was satisfactory. She married at 23. Her personality was mildly obsessional. She liked routine and tended to worry unnecessarily about trifles. Her interests were limited, but she was comparatively well adjusted and lived alone throughout heavy air raids relatively undisturbed. She denied any tendency to preoccupation with her health and compared her present lack of courage with her calmness at that time. In 1941 she was operated on for a retroverted uterus. Two years later she miscarried. A febrile illness with muscle and joint pains followed in 1945, necessitating three months in bed and a prolonged convalescence with frequent examinations of her heart, which she was told had been unaffected. An operation for a ruptured ectopic pregnancy was performed in 1946. She was very ill and required frequent blood transfusions. She experienced hot and cold sweats and felt she was going to die. After this she passed through a phase when she was nervous, irritable, unstable, and hypochondriacal. She recalled the sudden deaths of her two sisters and became preoccupied with the thought that she might be the third to die. She was not, however, aware of palpitation, and this episode of mental instability passed quickly. For the next eighteen months she kept reasonably well, but worried a good deal about the heart disease of her adopted daughter and her own inability to bear a child.

Home Conditions.—The relationship between the patient and her husband was satisfactory. There were no financial or special difficulties.

Course

The acute anxiety continued. She was anxious to co-operate in treatment and get well. Frequent and regular psychotherapeutic sessions took place, with only transitory improvement. The resting pulse rate rose to 120. Sedation with sodium amylbarbitone gave little relief. Her weight began to fall, and modified insulin therapy was used to prevent further weight loss, but no significant improvement followed this. She continued to have fears of death and episodes of panic. The tachycardia persisted and her general attitude to it remained unchanged.

In view of the persistent tachycardia, which was little influenced during sleep, specialist opinion was again sought and further electrocardiograms were done. The report stated that the heart was normal and the symptoms psychologically determined. The patient's mind became obsessed by an acute awareness of palpitation to the exclusion of all else. She was quite unable to accept or readjust herself to her tachycardia and became acutely agitated and hopeless in her outlook. A suicidal attempt followed. Electric-shock treatment was given because of these developments and a superficial improvement occurred; she became mildly euphoric and distractable. Careful examination, however, showed clearly that below this superficial veneer of well-being all the old symptoms continued. The tachycardia was unaffected. In two weeks a complete relapse occurred. A second course of shock treatment was ineffective. Under intravenous thiopentone her talk was restricted to the bumping of her heart, which she said she could not tolerate, and suicide, and how this could be accomplished.

At this stage she had been confined to bed for almost six months, and her illness was of 18 months' duration. All

treatment had been unsuccessful and the outlook seemed bad. In view of this and the intense self-awareness and severe mental tension, bilateral prefrontal leucotomy was recommended. Both the patient and her husband readily consented and the operation was performed on January 21, 1950.

Progress

First Week.—For two or three days after operation her memory and orientation were disorganized. She was disconnected in her talk and irrelevant in her replies. There was incontinence of urine for one week.

Second Week.—She got up and was usefully employed in simple ward duties and in the occupational therapy department, where she was especially encouraged. Striking features were the complete absence of tension, the calm facial expression, and the reduction of the pulse rate. The latter now ranged between 78 and 100 and averaged 85 when the patient was ambulant; for many months before operation it had varied between 110 and 150 and averaged 120 at rest.

Third Week.—She denied depression and suicidal thoughts, and never mentioned her previous awareness of her heart, though she was asked several times to describe the symptoms that worried her before operation. In reply to leading questions, however, she recalled that palpitation had worried her greatly and said that even now she had not got rid of it entirely. She was aware of it from time to time if she hurried, but it did not disturb her: she gave this information without evident anxiety as a circumstance to which she was indifferent. She was eager to get home. She was fully orientated and her memory steadily improved.

Fourth Week.—Her husband reported that she was better and happier than she had been for years. Her attitude to life was more matter-of-fact and unemotional. Her excessive anxiety, not only about herself but about a multiplicity of matters that used constantly to exercise her mind, was much diminished.

Fifth Week.—Her good progress continued. There was clearly emotional flattening, and special tests devised to assess objectively abstractive and learning ability showed considerable impairment, though she had been a woman of good average intelligence. Another finding of interest was a reduction in the basal metabolic rate to +6.

Seventh Week.—She was discharged from hospital and after a month's convalescence took up her household duties again; with these she coped adequately without help. Since discharge she has been seen regularly in the outpatient department and there has been no recurrence of her previous symptoms.

On January 2, 1951, a year after operation, she was reported to be well, coping with her duties, and showing no evidence of anxiety.

Comment

The salient features of this illness were severe persistent anxiety and tachycardia with raised basal metabolic rate. Diagnosis included a consideration of the relative importance of physical and emotional factors in producing and perpetuating the syndrome. Heart disease and thyroid disease were reasonably excluded within the limits of current knowledge. Basal readings of the metabolic rate are often difficult to obtain in the presence of severe mental tension, and Murray (1950) has drawn attention to the high figures that may be recorded in such cases in the absence of hyperthyroidism. When the syndrome was related to the facts of the history evidence of emotional factors emerged which made their nature meaningful. The reason for their continuance in the face of prolonged treatment remains obscure.

This case is representative of a group characterized by continued anxiety which responds disappointingly to all reasonable methods of treatment, although initially the

prognosis is considered to be good. Many, notwithstanding their burden of mental tension, can be kept usefully employed by supportive out-patient treatment with short periods in hospital during exacerbations. For some, however, their distress becomes so overwhelming and their functional efficiency so seriously impaired that they are obliged to remain at home or in hospital in a state of invalidism.

The patient described here became incapacitated in this way, and leucotomy afforded a striking and continued relief of symptoms. The general change in personality similar to that which has been described by others (Hoch, 1949) has not materially interfered with her social rehabilitation.

Summary

A case is described in which persistent anxiety, tachycardia and raised basal metabolic rate unresponsive to other methods of treatment were successfully treated by bilateral prefrontal leucotomy.

A year has passed without relapse.

The general change in personality has been compatible with excellent social rehabilitation.

The case demonstrates some problems in diagnosis and treatment.

This work was done at the Bristol Mental Hospitals, and I am obliged to Dr. R. E. Hemphill for affording me facilities to study this case. I am particularly indebted to Dr. Max Reiss for assessing thyroid activity with radioactive iodine, to Dr. E. C. Turton for estimating the basal metabolic rates, and to Dr. K. R. L. Hall for carrying out special psychological tests.

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CONGENITAL AFIBRINOGENAEMIA

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Bleeding, either spontaneous or due to trauma, may occur in a variety of conditions. One of these is due to a defect in the mechanism of blood-clotting. Hypotheses regarding the mechanisms involved in blood-clotting have changed during the last few years, but one concept that has not altered is the conversion of fibrinogen to fibrin by the enzyme thrombin. The following is a report of a case of congenital afibrinogenaemia.

Case Report

The patient was a male infant aged 11 months. His father was British and his mother of Eurasian extraction. The child was admitted on June 18, 1950, as an emergency for bleeding from the mouth. A lump had appeared on the right side of his face two weeks earlier and, as this would not resolve, the mother consulted her family doctor, who aspirated it from the inner aspect of the cheek. The bleeding continued for 12 hours, ceased after "an injection," but recurred. All attempts to arrest it failed, and the child was admitted to the Lady Ridgeway Hospital. In spite of treatment the bleeding continued for 24 hours after admission

and then ceased. The mother noticed small "blue" areas on various parts of the body—the right popliteal fossa and the left side of the chest—and she also stated that there was a tendency for three or four of these blue areas (each about the size of a halfpenny) to appear, especially on the chest and arms, whenever the child cut a tooth. With the eruption of the tooth the purpuric patches disappeared.

The child was full-term, the delivery was normal, and his birth weight was 5½ lb. (2.5 kg.). Umbilical haemorrhage began on the third day after birth and continued for seven days. This was quite copious and ceased after the local application of tincture of ferric chloride. When the child was 2 months old he had an attack of macroscopic haematuria for one night.

As the father had left Ceylon his family history is not available. There is no consanguinity of parents. One elder sibling, aged 4 years, gave no history of bleeding or any other illness.

The child was breast-fed for four months; a proprietary milk food was given for the next seven months. Orange juice in adequate quantities had been given from the age of 4 months. From the age of 7 months proprietary milk and cereal foods, calcium, and "abidec" drops had been administered. The infant could roll over at six months, but did not sit up at all.

On examination the child was extremely pale, with an icteric tinge of the skin; the tongue and lips were light pink in colour. A lymphangioma about the size of a lemon was present below the right lower jaw. There were a few localized areas of blue colour each about the size of a halfpenny over the left side of the chest, the right popliteal fossa, and the right tibial region. The small patch on the chest disappeared after a few days.

The child had an intermittent fever ranging from 100 to 102° F. (37.8 to 38.9° C.). He was given penicillin, 100,000 units three times a day for four days.

A blood count on June 24 showed: haemoglobin (5.4 g. Sahli), 31.7%; red cells, 1,820,000 per c.mm.; white cells, 7,800 per c.mm. (P. 42%, L. 58%); size of red cells, 7.8 µ (Pijper's halometer); bleeding time, 2½ minutes; clotting time, over 72 hours. On July 3 the marrow count showed: promyelocytes, 5%; myelocytes, 34%; metamyelocytes, 6%; polymorphs, 13%; eosinophil myelocytes, 3%; lymphocytes, 1%; megakaryocytes, 1%; normoblasts, 33%; proerythroblasts, 4%.

On July 6 a blood count showed: red cells, 1,820,000 per c.mm.; platelets, 60,000 per c.mm.; white cells, 25,400 per c.mm. (P. 27%, L. 73%, E. 0%); reticulocytes, 3%; haemoglobin, 35% (Dare's 5.6 g. %); colour index 0.96; bleeding time, 6½ minutes; clotting time, over 48 hours. Blood picture: anisocytosis, poikilocytosis, pessary forms, and polychromasia. Prothrombin time, over 24 hours (Quick's test, modified by Fullerton, 1940). (Control, 26 seconds.) Plasma proteins: albumin 3.48 g. %, globulin 2.22 g. %, fibrinogen nil—total 5.7 g. Tests for fibrinogen: (1) heating plasma at 60° C., no clot formation; (2) half saturation of plasma with NaCl, no precipitate. On the addition of "stypven" (Russell viper venom) no clot occurred even after 48 hours. The addition of 1 ml. of the child's blood to 0.5 ml. of normal blood resulted in clotting, suggesting that there was no antithrombic substance in the child's blood.

On July 7 the maternal grandmother's fibrinogen was 0.28 g. per 100 ml. and the mother's 0.21 g. per 100 ml. On July 10 the E.S.R. (micro-method) of the patient was 4 mm. at the end of one hour.

On July 8 the child was given a blood transfusion of 180 ml. of whole blood. There was a severe reaction, the temperature rising to 107° F. (41.7° C.), and the transfusion was stopped. Owing to technical difficulties the transfusion was given into the internal saphenous vein after exposure. The superficial wound was sutured and the vein clamped at two points and ligated. Oozing continued from the margins of the incision, and the application of adrenaline