"Silent" and puzzling cases of heart failure have been ascribed to endomyocardial fibrosis (Ball, Williams, and Davies, 1954), a condition almost confined to tropical Africa. This takes the place of coronary disease as the prime cause of cardiac death in adults, and the aetiology is quite unknown.

The distribution of coeliac disease and fibrocystic disease provides some unsolved puzzles. Coeliac disease is said not to occur in parts of the world where wheat is not a regular item of diet (Sheldon, 1955). In Uganda, however, bread is eaten occasionally in large parts of the country, but one wonders how the disease would be recognized when fat is virtually absent from the diet. Possibly some cases are being missed. The diagnosis of fibrocystic disease is made difficult because the pancreatic enzymes are absent in acute kwashiorkor and slowly return with improvement. One possible case was seen but could not be followed up.

These few scattered examples can only hint at a subject of great importance and interest, the study of which will undoubtedly provide valuable clues to the aetiology of disease, both temperate and tropical.

Summary

An attempt has been made to describe some common and urgent tropical paediatric problems as they were encountered during a two-year attachment to the Uganda Medical Service, together with some of the ethnographical, economic, and human background. Even for this very limited purpose, two years is but a fleeting acquaintance, and there will undoubtedly be omissions and distortions.

Infant mortality is very high and there is a high death rate throughout childhood, the main causes being malnutrition, malaria, respiratory disease, gastro-enteritis, helminthiasis, and sickle-cell anaemia. Medical work is hampered largely by financial stringency, and to a less extent by superstition and suspicion on the part of patients.

The conception of a tropical branch of paediatrics is justified by the different pattern of child life and disease, both temperate and specifically tropical, and the subject has received little prominence in medical teaching, considering the huge child populations of the tropics and subtropics.

A study of the differential geographical distribution of disease will undoubtedly throw much light upon aetiology, and is worthy of greater effort.

It is a pleasure to express my thanks to the Board of Governors of the Hospital for Sick Children, Great Ormond Street, for having made possible the experience underlying this paper. I wish also to thank Dr. Hugh Trowell for his constant help and encouragement in Uganda, and Professor Alan Moncrieff and Dr. Wilfrid Sheldon, who kindly offered comments on and criticisms of this paper.

REFERENCES

- Ball, J. D., Williams, A. W., and Davies, J. N. P. (1954). *Lancet*, **1**, 1049.
 Blankhart, D. M. (1953). *Docum. Med. geogr. trop. (Amst.)*, **5**, 7.
 Jelliffe, D. B. (1952a). *Trans. roy. Soc. trop. Med. Hyg.*, **46**, 13.
 — (1952b). *Pediatrics*, **9**, 544.
 — (1955). *Ibid.*, **16**, 398.
 Luder, J. (1956). *E. Afr. med. J.*, **33**, 100.
 Sheldon, W. P. H. (1955). *Lancet*, **2**, 1097.
 Trowell, H. C. (1956). *Trop. Dis. Bull.*, **53**, 121.
 — Davies, J. N. P., and Dean, R. F. A. (1954). *Kwashiorkor*. London.
 Welbourn, H. F. (1955). *J. trop. Pediat.*, **1**, 34, 98, 161.
 Wilson, A. M. M., and Luder, J. (1957). In press.
 Woodruff, A. W. (1955). *Brit. med. J.*, **1**, 1297.
 Zourendyk, A. (1947). *S. Afr. J. med. Sci.*, **12**, 167.

THE NATURAL HISTORY OF POLYARTERITIS*

BY

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In 1952 the Collagen Diseases and Hypersensitivity Panel of the Medical Research Council began a clinical trial of cortisone in polyarteritis nodosa. It was clearly unjustifiable to have concurrent untreated controls, and so some form of retrospective control was necessary. Cases reported in the literature are unsuitable because they are a selected group and are often inadequately followed up. It was thus necessary to collect a new body of data from an unselected and consecutive series of cases studied specially for this purpose. Such a study presented an opportunity not only to learn the prognosis and duration of the disease but also to make a comprehensive survey of its natural history and pathology. Some of the special aspects of this investigation have been reported already (Rose, 1956, 1957; Rose and Spencer, 1957); in the present paper the whole of the results are presented in summary form. The complete report and the case histories are lodged with the Medical Research Council, from whom a copy may be borrowed on request.

The survey was based on all histologically proved cases of polyarteritis under care at nine teaching centres during the period 1946 to mid-1953; 111 cases were obtained. Of these, 90 were located from diagnostic indices, and the remaining 21 by a search of all the necropsy reports (and sometimes the sections also) for the period in question. In any possible cases of polyarteritis all the available sections were studied by one or other member of the panel of pathologists set up for this purpose by the Medical Research Council.‡ Doubtful cases were seen by the whole panel. Full reports were provided on all approved cases, and these form the basis of the histological analysis in the present report. The panel did not, however, take responsibility for the analysis.

There was evidence that the number of cases diagnosed was tending to increase during the period of the survey, particularly among the elderly. This may well have represented a true increase in the incidence of the disease; but the limitations of a retrospective survey prevent any firm conclusions. Unfortunately no trend was apparent towards either earlier diagnosis or more frequent ante-mortem diagnosis.

Classification

In a preliminary survey of data it appeared that some classification might usefully be based on the presence or absence of pulmonary involvement. Unfortunately adequate histological material from the lungs was not always available, and the assignment of patients to one or other group was therefore based in addition on other features which were

*A summary of a report to the Collagen Diseases and Hypersensitivity Panel of the Medical Research Council.

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found to be characteristic of cases with demonstrable pulmonary polyarteritis. By this means a group of cases was obtained distinguished by the presence in each case of at least three (usually more) of the following features: a characteristic respiratory illness preceding the onset of systemic polyarteritis; high blood eosinophilia (1,500 or more per c.mm.); numerous eosinophils in acute polyarteritic lesions; pulmonary polyarteritis; granulomatous polyarteritis; giant cells in polyarteritic lesions; and the presence in viscera of necrotizing or granulomatous lesions not demonstrably related to arteries. The term "polyarteritis nodosa with lung involvement" is suggested for this group, which comprised one-third of the cases.

From a study of previous reports it appears that similar cases have been given a variety of names, including polyarteritis nodosa, periarteritis nodosa, Wegener's granuloma (Godman and Churg, 1954, after Wegener, 1936), Loeffler's syndrome (Buckles and Lawless, 1950), Harkavy's syndrome (after Harkavy, 1941), "giant-cell granuloma associated with lesions resembling polyarteritis nodosa" (Howells and Friedmann, 1950), "periarteritis nodosa in granuloma of unknown aetiology" (Weinberg, 1946), "necrotizing granulomatosis and angiitis" (Fienberg, 1953), "diffuse eosinophilic arteritis" (Lumb, 1950), "allergic granulomatosis, allergic angiitis, and periarteritis nodosa" (Churg and Strauss, 1951), etc.

The reasons for adding yet another name to this list are, first, that a newly defined group of cases is being described, and, secondly, that the terms listed above are mostly too cumbersome for general use. The term "polyarteritis nodosa with lung involvement" is suggested because it stresses what appears to be the chief criterion of the syndrome, and it does not imply any particular aetiology.

The characteristic features of the group with lung involvement were all either completely absent or rare in the other groups. Microscopically, however, the majority of lesions in the two groups were often indistinguishable; that is to say, the characteristic lesions in cases with lung involvement were often small in number. It is not clear whether these two syndromes are distinct diseases or merely variants; but it is suggested that this is the most valid classification so far proposed, and that it is of practical value in clarifying the natural history of the disease. Unfortunately it appears that steroid treatment makes it more difficult to discover at necropsy the characteristic lesions associated with lung involvement.

In six cases the available evidence was suggestive of lung involvement, but not conclusive. These cases are described separately, as also are seven other cases which differed greatly from both the two main groups.

Polyarteritis Nodosa Without Lung Involvement

There were 66 patients in this group (41 males and 25 females); 55 of them died and 54 came to necropsy. The age incidence showed a steady rise throughout life to a maximum in the seventh decade. The onset of the disease most often fell in the winter months. No special trends were apparent with regard to the patients' occupations or smoking habits.

At the time of onset of the disease 18 patients (27%) had chronic respiratory infections (bronchiectasis, chronic bronchitis, or suppurative otitis media), and another 12 (18%) had had recent acute upper respiratory infections (attributable in at least eight to haemolytic streptococci); at least three of the patients with recent acute infections had not received any drugs. In six patients (9%) there was active or quiescent polyarthritides indistinguishable from rheumatoid arthritis.

A constitutional illness (with loss of weight, fever, and tachycardia) was prominent at some stage of the disease in nearly all patients. Erythrocyte sedimentation rates were almost always high, and anaemia and leucocytosis were very common; slight or moderate eosinophilia (>400 per c.mm.)

was noted at some stage in 30% of patients. Measurement of the serum protein usually showed increased globulin and lowered albumin levels.

There was evidence of coronary polyarteritis in 32 patients (48%). The commonest clinical manifestations were cardiac failure and enlargement; cardiac pain was uncommon, although necropsy often revealed infarcts (usually microscopic).

Pulmonary manifestations in this group of patients were all attributable to infections or heart failure; there was no evidence of any specific lesions.

Renal involvement occurred at some stage in 52 patients (79%). The commonest lesion was renal polyarteritis, present as the only renal lesion at necropsy in 39 cases. Clinically this was associated in nearly all cases with proteinuria and microscopic haematuria, and sometimes also with a few granular and hyaline casts; 10 of these patients died of renal failure, and at necropsy they showed infarcts (often extensive) in the renal cortex.

A specific form of glomerulitis was present as the only renal lesion at necropsy in 16 cases. This was characterized in its initial phase by capillary microthrombi, focal fibrinoid necroses, polymorph infiltration, and capsular proliferation (often with crescent formation); clinically such cases showed macroscopic or heavy microscopic haematuria and early renal failure. Hypertension was not a feature of this initial phase. The three patients who survived this stage developed progressive hypertension and uraemia, and died within a year; at necropsy the microscopical appearances resembled those of ordinary chronic glomerulonephritis. In six additional cases the typical acute glomerulitis was associated with renal polyarteritis.

Significance of Raised Blood Pressure

In assessing the significance of an elevated blood pressure, the average normal pressure for each patient's age and sex was calculated from the tables compiled by Hamilton, Pickering, Roberts, and Sowry (1954). If the systolic or diastolic pressure exceeded these values by 30 or 20 mm. Hg respectively, the patient is referred to as having hypertension. Alternatively, where during the period of observation the systolic or diastolic pressure rose by 30 or 20 mm. (or more) respectively above the initial value the patient is said to have developed hypertension.

Valid records of arterial blood pressure were available in 55 cases. In 24 the pressure was normal on all occasions; at necropsy in these cases recent arterial and glomerular lesions were common, whereas healed lesions with associated ischaemic changes were seen only once. In 15 cases hypertension was observed to develop during the course of the disease; urinary evidence of renal lesions preceded any rise in pressure in all cases where data were adequate. At necropsy 12 of them had either healed renal polyarteritis or chronic glomerulitis; the other three had ischaemic changes in the kidneys not attributable to arteriosclerosis, but without positive evidence of polyarteritis. In 16 cases the blood pressure was raised at the initial measurement; but in 14 of these this measurement was made several months after the onset of the disease, and only in the other two was there definite evidence that hypertension preceded polyarteritis nodosa. Furthermore, in eight cases in this group the hypertension was found to be advancing rapidly during the period of observation, and at necropsy all but 2 of the 16 cases had healed renal polyarteritis. It is concluded that hypertension in polyarteritis nodosa is in almost all cases a sequel of renal polyarteritis or glomerulitis, but that the pressure rises only during the healing or healed stages of these lesions. Once initiated, hypertension is progressive; it terminated in the malignant phase in at least 11 patients in this series (10 males and 1 female). At necropsy the severity and extent of polyarteritic lesions were approximately the same in cases with normal and cases with high blood pressure; healed lesions were more frequent in the latter.

Symptoms and Signs

There was evidence of gastro-intestinal polyarteritis in 46 patients (70%). The commonest symptom was poorly localized pain; haemorrhage was common, and sometimes severe. Physical signs were usually slight or absent. At necropsy submucous haemorrhages and superficial ulcers were frequent. Polyarteritic lesions were usually confined to the terminal branches of the mesenteric arteries.

The liver was involved in at least 29 out of 54 necropsy cases (54%), but clinical evidence was usually lacking. Pathologically, the lesions appeared to excite more surrounding fibrosis than in other organs. In two patients necropsy showed diffuse hepatitis; neither had demonstrable hepatic polyarteritis.

Muscle pain and tenderness were frequent (often initial) manifestations of the disease, but the presence of nodules was recorded only once. Muscle was taken for biopsy on 26 occasions, on 12 (46%) of which polyarteritic lesions were demonstrated; in several of the negative cases the amount of muscle removed was very small. The impression was gained that the pectoral muscles were probably the site of choice for biopsy.

Arthritis arose during the course of the disease in 18 patients (27%). In six of these the changes were acute and transient (sometimes migratory, as in rheumatic fever). In eight others they were chronic and more closely resembled a rheumatoid type. Hypertrophic osteoarthropathy developed in two patients, one of whom had no pulmonary lesions of any sort.

The spleen became palpable in eight patients (in two of whom sections taken at necropsy did not show any splenic polyarteritis). Splenic polyarteritis (usually in arterioles or small trabecular arteries) was seen in 16 of the 39 cases from which sections were available.

There was clinical evidence of peripheral nerve lesions in 24 patients (36%). Combined sensory and motor loss was usually present. In seven patients there was clear evidence of lesions of isolated peripheral nerve trunks; in the remainder the signs were those of a more widespread (but still asymmetrical) peripheral neuritis. Sections of nerve were available from 12 of these cases, in seven of which polyarteritis was seen in the vasa nervorum. It is suggested that all peripheral nerve lesions in polyarteritis nodosa are due to local polyarteritis.

Lesions of the central nervous system were in most cases attributable to hypertension or atheroma. Microscopical proof of cerebral polyarteritis was found in only two cases; but in nine others there was no clear explanation for the clinical evidence of cerebral damage, and local polyarteritis cannot be excluded as a possible cause.

Focal indurated skin lesions, probably all due to local polyarteritis, occurred in 18 patients (27%). These ranged from subcutaneous nodules to superficial papules, often with haemorrhage and ulceration. A characteristic surrounding purple reticulation was a common sequel. Biopsy of the lesions was undertaken on 15 occasions, on 10 of which there was demonstrable local polyarteritis; in the negative sections the presence of infarction or infection usually made interpretation impossible. Other skin lesions included purpuric rashes, painful subcutaneous oedema, and (in one case) scleroderma.

At necropsy complete absence of any active lesions was found in nine cases (in three of which cortisone had been given). In 29 cases polyarteritic lesions were visible to the naked eye; aneurysms (usually few in number) occurred in 10 cases. It was not possible to base any distinct classification on the size of artery involved.

A correct clinical diagnosis was reached in 31 patients (47%), but was frequently delayed; 22 of these were proved by biopsy. In retrospect it appears that a correct clinical diagnosis could have been achieved on the basis of the recorded data in 47 patients (71%). Thus, despite its varied clinical manifestations, polyarteritis nodosa does, nevertheless, produce recognizable clinical patterns in most cases.

Prognosis

Of the 66 patients in this group, 12 were treated with cortisone or corticotrophin. Of the remaining 54, 51 died (one-third within three months and two-thirds within six months); three cases remitted fully, in two of which the lesions appeared to be confined to the skin and joints. Renal lesions were the cause of death in 65% of cases; the immediate cause was uraemia in those who died early, but hypertension in the majority of late deaths. In the remaining cases death was due mainly to coronary or gastro-intestinal polyarteritis. The severe constitutional illness contributed to death in most instances.

These figures on the prognosis of the disease are heavily weighted by cases that were first diagnosed at necropsy. A true assessment of the prognosis of the untreated disease needs to be based on cases proved in life (by biopsy) during the pre-cortisone era. In order to increase the small number of such cases available, the survey was extended to five additional teaching hospitals, covering the period 1941 to mid-1950—that is, prior to the introduction of cortisone and corticotrophin. In all, 16 cases were obtained; these have been described in detail elsewhere (Medical Research Council, 1957). Of the 16, 11 died (all but one within a year), in two the disease was still active (8 and 1½ years respectively from the onset), and three remitted. Of the latter, only one had evidence (based on macroscopic haematuria) of visceral lesions; this case remitted after three months. In the other two cases lesions were apparently confined to skin and joints, and remission occurred after five and seven years. The period of follow-up was three, three, and five years respectively from the date of remission.

Polyarteritis Nodosa with Lung Involvement

There were 32 cases in this group (16 males and 16 females). In eight patients (25%) there was a long history of respiratory infection preceding (so far as could be ascertained) the onset of polyarteritis. A past history of rheumatic fever was obtained from five patients, and one other was found at necropsy to have mitral stenosis (a total of 19%). One patient had rheumatoid arthritis.

Loss of weight, fever, and tachycardia were prominent features. Eosinophilia was observed in 69% of the cases from which data were available; it exceeded 1,500 per c.mm. at some stage in 50%, but was often intermittent. In 36% the highest count was 5,000 per c.mm. or more.

A respiratory illness occurred in all patients, and in all but one it preceded the onset of systemic polyarteritis (usually by less than one year, but occasionally by up to seven years). The evidence suggests that this phase of the illness was associated with the presence of specific pulmonary lesions.

Clinically the main features (Rose, 1957; Rose and Spencer, 1957) were those of asthma, pneumonia, or chronic bronchitis. The asthma appeared to be distinct from ordinary allergic asthma in that there was no family history, symptoms started at any age, extremely high eosinophilia was frequent, and necropsy usually revealed necrotizing lesions and polyarteritis in the lungs. The pneumonia ranged in clinical severity from transient symptomless infiltrations to large chronic abscesses; there was no response to antibiotics.

Haemolytic streptococci were isolated at some stage from 23% of those patients whose sputum was cultured; tubercle bacilli were never found. Radiological examination revealed a very variable pattern of consolidation. At necropsy (Spencer, 1957) severe lung damage was present in nearly all cases. The commonest change observed (50% of cases) was the presence of nodular or caseous lesions, often resembling tuberculosis (but with a complete absence of tubercle bacilli). Other lesions included haemorrhagic pneumonia, cavities, infarcts, fibrosis, and bronchiectasis. Microscopy showed characteristic necrotizing or granulomatous lesions in 61% of cases, and pulmonary polyarteritis

in 14 of the 20 cases from which adequate sections were available; diffuse and focal eosinophil infiltration was frequent.

In five cases granulomatous lesions occurred in the nose or middle ear. This lesion seems to be confined to cases with lung involvement.

The incidence of polyarteritis in other organs and the accompanying clinical manifestations were in the main similar to those seen in cases without lung involvement. Hypertension was less frequent; this appeared to be related to the shorter survival of cases with lung involvement once systemic polyarteritis was established. In the alimentary tract superficial ulceration (at all levels, from mouth to colon) tended to be more frequent and extensive than in cases without lung involvement. Necrotizing or granulomatous lesions not demonstrably related to arteries were encountered in the liver, kidneys, spleen, lymph nodes, and heart. Foci of collagen necrosis were seen very occasionally in the splenic capsule and trabeculae, and in the endocardium. In patients who had a high blood eosinophilia at the time of death there was usually a widespread diffuse and focal infiltration of interstitial tissues with eosinophils.

Of the 32 cases, eight received cortisone or corticotrophin. Only one patient (a cortisone-treated case) survives. The mean total duration of the disease was rather longer than in patients in whom the lungs were spared, but the period of survival from the onset of systemic polyarteritis was less. The various characteristic forms of pulmonary disease accounted for 42% of all deaths; another 26% were due to renal lesions (mostly acute).

Only four cases were obtained that had been proved by biopsy in the pre-cortisone era. Three of these patients died (2, 8, and 18 months respectively from the onset of the disease); in the surviving case the disease was still slightly active 16 years from its onset.

Aetiology

The aetiology of the disease remains obscure. The most likely clue is the relation to preceding chronic and acute respiratory infections (especially those due to haemolytic streptococci). One of the chief problems is to distinguish whether the significant association is with the infections or with their treatment. The evidence from the present survey favours the incrimination of the infection itself, although no definite conclusions are justified (Rose, 1956; Rose and Spencer, 1957). The important observations are the association of some cases with haemolytic streptococcal infections not treated by any drugs, the association of the other cases with rheumatic fever, and the prolonged course of the disease with exacerbations and partial remissions seemingly unrelated to any drug therapy.

Other Examples of Polyarteritis

Six patients showed two of the features listed above as characteristic of polyarteritis nodosa with lung involvement, but this was considered insufficient evidence to warrant their inclusion as definite examples of the syndrome. It is possible that their classification would have been simpler if more adequate data had been available. One of these cases was of special interest because of its association with acute rheumatic fever.

Seven other cases were classified separately because they differed markedly from both of the two main groups. They have been regarded (with the possible exception of the first two cases described below) as examples of separate diseases which happen to show a similar histological lesion. They were:

1. A case of typical disseminated lupus erythematosus with terminal polyarteritis of mesenteric and peria renal arteries and a vas nervi. An association between these two diseases has been reported previously by Karani (1953).
2. A case of dermatomyositis in which necropsy revealed localized polyarteritis in voluntary muscle and in the oesophagus.
3. A case of scleroderma with cystic lung changes in which polyarteritis was confined to the areas of damaged lung.

4. A case of fibrosarcomatosis with terminal polyarteritis in skin and voluntary muscle.

5. and 6. Two cases of polyarteritis confined to branches of the pulmonary artery, occurring in patients with pulmonary hypertension (due in one case to mitral stenosis and in the other to emphysema). At least 10 cases of this sort have been described previously (Jones, 1942; Old and Russell, 1950; McKeown, 1952; Symmers, 1952; Hicks, 1953; Wade and Ball, 1957), and Parker and Weiss (1936) described five cases of mitral stenosis in which there was necrosis of pulmonary arterioles without any inflammatory reaction. The condition seems in some respects comparable to the systemic arteriolar necroses of malignant hypertension.

7. An infant who died at the age of 4 months, in whom necropsy revealed multiple aneurysms on the aorta and its branches and on the coronary arteries. Histologically the process seemed to have started as a necrotizing endarteritis. There was no clinical or pathological evidence of bacterial or mycotic infection.

Conclusions

This investigation offered a unique opportunity to assemble information on a large and unselected series of cases of polyarteritis. Certain features have emerged from it which are new or which now receive more detailed support.

1. The suggested classification into two groups, according to the presence or absence of lung involvement, appears to be the most reasonable so far available. It stresses the peculiar importance of lung involvement in the disease: for it is remarkable that lung lesions, if they occur at all, almost always precede lesions in other organs (sometimes by years), and that they are commonly associated with characteristic lesions in other organs of a type not seen in cases without lung involvement. These characteristic lesions may be few in number, and it is not certain whether these two syndromes represent distinct diseases or merely variants of the same condition.

2. The relation between the onset of the disease and preceding respiratory infections has been noted before—for example, Grant (1940) and Miller and Daley (1946)—but it now receives further support. An association with rheumatoid arthritis and with rheumatic fever is also apparent.

3. It has been suggested by Knowles, Zeek, and Blankenhorn (1953) that hypertension is a common cause of polyarteritis nodosa. The more detailed evidence now available shows that hypertension in polyarteritis nodosa is almost always a result of renal involvement (either arterial or glomerular). It is of interest that the arterial pressure is normal during the acute stage of these lesions, and begins to rise only when healing and fibrosis occur.

4. The existence is confirmed of a characteristic form of glomerulitis peculiar to cases of polyarteritis nodosa, as described by Davson, Ball, and Platt (1948). It is shown that some patients with this lesion survive into a chronic phase, in which they develop hypertension and renal failure.

5. Only 20 cases were located which had been proved by biopsy in the pre-cortisone era. But, although small, this group provides the largest consecutive series so far available for the assessment of the course and prognosis of the untreated disease.

Summary

The clinical and pathological findings are presented from a series of 111 proved cases of polyarteritis collected in nine teaching hospitals from the period 1946 to mid-1953. A classification is suggested and some evidence relating to the aetiology is discussed.

I am indebted to Professor P. R. H. Lovell and Professor Sir George Pickering, who gave invaluable advice at every stage of the work; also to Professor G. Payling Wright, Professor C. V. Harrison, and Dr. A. H. T. Robb-Smith. I thank Dr. H. Spencer, who undertook the greater part of the extensive histological work.

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atism); Guy's Hospital; Hammersmith Hospital and Postgraduate Medical School of London; London Hospital; Radcliffe Infirmary, Oxford; Royal Infirmary, Edinburgh, and the hospitals served by the Edinburgh University Department of Pathology; Royal Infirmary, Manchester; St. Mary's Hospital; St. Thomas's Hospital. The following hospitals co-operated in a limited extension of the survey to include more cases proved by biopsy: Middlesex Hospital; Royal Victoria Infirmary, Newcastle upon Tyne; St. Bartholomew's Hospital; University College Hospital; Westminster Hospital.

REFERENCES

- Buckles, M. G., and Lawless, E. C. (1950). *Dis. Chest*, **18**, 312.
 Churg, J., and Strauss, L. (1951). *Amer. J. Path.*, **27**, 277.
 Davson, J., Ball, J., and Platt, R. (1948). *Quart. J. Med.*, **17**, 175.
 Fienberg, R. (1953). *Amer. J. Path.*, **29**, 913.
 Godman, G. C., and Churg, J. (1954). *A.M.A. Arch. Path.*, **58**, 533.
 Grant, R. T. (1940). *Clin. Sci.*, **4**, 245.
 Hamilton, M., Pickering, G. W., Roberts, J. A. F., and Sowry, G. S. C. (1954). *Clin. Sci.*, **13**, 11, 37.
 Harkavy, J. (1941). *Arch. intern. Med.*, **67**, 709.
 Hicks, J. D. (1953). *J. Path. Bact.*, **65**, 333.
 Howells, G. H., and Friedmann, I. (1950). *J. clin. Path.*, **3**, 220.
 Jones, G. M. (1942). *Ann. intern. Med.*, **16**, 920.
 Karani, S. (1953). *Postgrad. med. J.*, **29**, 321.
 Knowles, H. C., Zeek, P. M., and Blankenhorn, M. A. (1953). *A.M.A. Arch. intern. Med.*, **92**, 789.
 Lumb, G. (1950). *Brit. med. J.*, **2**, 645.
 McKeown, F. (1952). *Brit. Heart J.*, **14**, 25.
 Medical Research Council (1957). *Brit. med. J.*, **1**, 608.
 Miller, H. G., and Daley, R. (1946). *Quart. J. Med.*, **15**, 255.
 Old, J. W., and Russell, W. O. (1950). *Amer. J. Path.*, **26**, 789.
 Parker, F., and Weiss, S. (1936). *Ibid.*, **12**, 573.
 Rose, G. A. (1956). *Proc. roy. Soc. Med.*, **49**, 289.
 — (1957). *Brit. J. Tuberc.*, **51**, 113.
 — and Spencer, H. (1957). *Quart. J. Med.*, **26**, 43.
 Spencer, H. (1957). *Brit. J. Tuberc.*, **51**, 123.
 Symmers, W. St. C. (1952). *J. clin. Path.*, **5**, 36.
 Wade, G., and Ball, J. (1957). *Quart. J. Med.*, **26**, 83.
 Wegener, F. (1936). *Verh. dtsch. path. Ges.*, **29**, 202.
 Weinberg, T. (1946). *Amer. J. clin. Path.*, **16**, 784.

NEUROPSYCHIATRIC SEQUELAE OF LIGHTNING STROKE

BY

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On July 14, 1955, lightning struck the racecourse at Ascot, killing two people and injuring a number of others. 51 were taken to various casualty centres in the vicinity, the majority suffering apparently from a minor degree of shock only; 31 were not detained. Twelve persons were rendered unconscious for more than a few moments, and two of these died from head injuries. The immediate effects, apart from shock, were in the nature of burns, temporary paralyses of the limbs, paraesthesiae, tremors, transient deafness and dysphasia, numbness, and headache (Arden *et al.*, 1956).

Injury or death from lightning is not a rarity in this country. It causes about five to ten deaths annually in England and Wales according to Hughes (1956), the victims usually being the unfortunate individual, or small group, trying in vain to shelter from the storm. Luckily, incidents on the scale of the one at Ascot occur very rarely. There was the well-known Koenigsberg disaster in 1925, which was fully reported by Panse (1925). Much more recently a lightning incident occurred at Aldershot.

Much of the relevant medical literature is concerned with the immediate effects of lightning injury. Neuropsychiatric sequelae are occasionally mentioned, but it is difficult to get a clear idea of their incidence. Psychiatric effects are recorded by Critchley (1932), the

principal ones being paranoid states and chronic psychoneurotic illnesses. Arden *et al.* reported that a few of the patients struck at Ascot had residual emotional disturbances, but no details were given. Hysterical manifestations after injury by lightning were also noted by Critchley (1932), particularly functional blindness, deafness, and loss of speech, and Langworthy (1936) mentioned hysterical aphonia as an occasional late sequel of injury by electricity. Hysterical symptoms were found by Critchley (1934) to be more common in lightning than in electrical injury. He also described a heterogeneous group of neuropsychiatric sequelae, which included transient pains, vasomotor disturbances, and spasms occurring in the limbs. A case of arterial spasm in the right arm and leg with transient paralysis has been reported by Currens (1945).

In the frequent references in the literature to paraesthesiae and loss of power in the limbs as the result of lightning stroke, opinions differ concerning the aetiology, although most authors emphasize a possible organic cause (Critchley, 1932; Cambier, 1953). Bach (1950) claimed to have demonstrated the organic nature of so-called neurotic symptoms following trauma by lightning. In one of his cases a demonstrable left-sided internal hydrocephalus was regarded as responsible for a fairly typical picture of post-traumatic neurosis, which had supervened in a man aged 38 who had been unconscious for two hours as a result of lightning stroke, and had later developed a temporary paralysis and dysphagia. A second man who showed symptoms of cerebral irritation became depressed, paranoid, and hypochondriacal, and later committed suicide. Haemorrhages in the occipital cortex were seen and reference was made to other similar cases in the German literature, and also to the findings described by Hassin in the brains of electrocuted criminals. Among the more clearly defined neurological sequelae are several references to cases of paralysis of both upper and lower motor neurone types (Critchley, 1934; Langworthy, 1936), and cases presenting as amyotrophic lateral sclerosis are described (Leys, 1942; Woods, 1952). Sphincter disturbance appears to be uncommon. A variety of ocular changes have been reported by Critchley and others, of which cataract seems to be the most common. Abnormal E.E.G. recordings have been noted; Cambier has reported minor changes in the theta rhythm.

The Inquiry

In view of the diversity of neuropsychiatric sequelae described, as well as their uncertain incidence, it seemed desirable, as Panse (1925) urged long ago, to inquire further into the consequences of lightning stroke. In describing the Koenigsberg incident of 1925, when three were killed and 26 injured as a result of lightning stroke, Panse referred to four cases (16%) as having reacted "psychopathically" to the trauma. Fear and anxiety were the common immediate effects. In no case did a traumatic neurosis ensue.

In the recent Aldershot incident a party of men were struck by lightning, and one man was killed. There were five other casualties, and medical reports were received relating to four of them. A month after the incident two men were still disabled by psychiatric symptoms. One, who had earlier displayed signs of cerebral irritation, had developed a mild anxiety state with loss of weight, poor appetite and sleep, tremors, apprehension, and dizzy spells; and the second, who was a close friend of the man killed in the incident, has continued to complain of weakness of the legs, dizzy spells, and headache, symptoms which are regarded as hysterical in nature.