bilateral cataract, which is by no means uncommon at his age. It is, however, possible that reduced blood supply and age were complementary factors in the opacification of the lenses. Ophthalmoscopic assessment of the condition of retinal blood vessels was not possible in the presence of the cataracts.

A rather intriguing point is how the cerebral blood supply was maintained. Obviously the supply through the left common carotid artery was completely cut off. Although the right common carotid was patent it was not in communication with the main current of aortic blood, since the innominate artery was totally occluded. There was no expansile pulsation in it. We may be justified in assuming that the process of occlusion was slow enough to allow collateral channels to open up adequately through whatever communication was available. The only other arteries that can carry sufficient blood to the brain are the vertebrals. The left subclavian artery was completely occluded as far as it could be removed. Whether the thrombus extended and involved the left vertebral artery is unfortunately not known. The lumen of the right subclavian artery was patent. It seems reasonable to conclude, in spite of the small material evidence, that anastomotic channels were established between the intercostals, internal mammary, and lateral thoracic arteries which fed the right subclavian, common carotid, and vertebral arteries.

The presence of tortuous pulsatile arteries on the chest wall evidently lends further support to such a conclusion. From the existence of similar arteries on the abdominal wall it appears that these collaterals were also reinforced with a supply from the external iliac and femoral arteries. While this can also explain the circulation in the right upper limb, it is not easy to suggest any possible channel of supply to the left, which was also apparently well nourished and warm. It would certainly be interesting to know if the left vertebral artery, and the subclavian artery beyond it, were patent.

The popliteal blood pressure was recorded with a standard arm cuff, and due allowance for this should be made. But the high pulse pressure is striking. It is probably a common expression of the extensive collateral anastomosis and atherosclerosis.

Unfortunately a complete post-mortem examination was not possible. In compliance with the sentiments of the patient's relations we were obliged to content ourselves with removing only the aortic arch with the proximal parts of its branches and a small piece of the right lung. From the clinical and fluoroscopic findings it was thought that the mouths of the three branches were blocked with atheromatous lesions, which are known to occur predominantly round the ostia of aortic branches. The necropsy findings of the big ante-mortem thrombus were very much unexpected. Thrombus formation in an aneurysmal sac is not very uncommon. But in its absence it is surprising how such a big clot can form in the arch where the turbulence of blood is very great indeed. Ross and McKusick (1953) found only three examples in 44 patients, and in all of them the thrombus was in syphilitic aneurysmal sacs.

In our case the histological picture of an atheroma of the aorta was unmistakable. Primary atheroma of the aortic arch is rare. Ross and McKusick think that trauma may play some part in the pathogenesis of atheroma in the arch. Our patient gave no history of physical violence on the chest, at least of any severe degree which he could remember. Atheromatous changes can occur on pre-existing syphilitic aortitis. There were no pathological changes suggestive of such a lesion in the media and adventitia. Furthermore, there was no history of syphilis and the blood Wassermann reaction was negative. The only abnormality found in the left subclavian, left common carotid, and innominate arteries was the presence in them of organizing and recanalizing thrombi. There was absolutely no pathological lesion in their walls. It may naturally be concluded that the primary atheromatous ulcer on the wall of the aortic arch helped in the formation of the thrombus which gradually increased in size and extended into all the three branches of the arch of the aorta.

Summary

A case of aortic arch syndrome characterized by absence of pulsation in all the arteries of the upper limbs and neck, with high systolic blood pressure in the vigorously pulsatile arteries of the lower limbs and evidence of collateral circulation on the abdominal and chest walls, is described. From its resemblance to the clinical syndrome of coarctation of the aorta it has been called reverse coarctation syndrome. The patient died as a result of associated bronchopneumonia.

The patient had very few symptoms and physical signs referable to impaired blood supply to the head and neck and to the upper limbs. The possible collateral channel of supply is discussed.

Only a limited necropsy study was possible. There was extensive atheroma with calcification in the wall of the aortic arch. The upper third of its lumen was occupied by an ante-mortem thrombus which extended into and completely obliterated the lumina of the innominate artery, and the left common carotid and subclavian arteries as far as they could be removed. The right common carotid and subclavian arteries were patent. Histologically, organizing and recanalizing thrombi were found in the occluded arteries. Except atheroma in the wall of the aortic arch no other pathological lesion was found in the walls of any of the arteries.

We are grateful to Dr. M. N. Sarkar, superintendent, Calcutta National Medical College Hospital, and to Dr. J. C. Saha for permission to publish this case. We thank Professor A. C. Guha, Professor S. N. De, and Dr. Shyamal Sen for their help in the pathological studies; and Dr. Amal Ghosal and Dr. Subhas Mukherjee for their valuable assistance in the study of the case.

REFERENCES

- Barker, N. W., and Edwards, J. E. (1955). *Circulation*, 11, 486.
 Caccamise, W. C., and Whitman, J. F. (1952). *Amer. Heart J.*, 44, 629.
 Frovig, A. G. (1946). *Acta psychiat. Kbh.*, Supp. 39.
 Harbitz, F. (1926). *Arch. Path. Lab. Med.*, 1, 499.
 Marinesco, G., and Kreindler, A. (1936). *Presse méd.*, 44, 833.
 Ross, R. S., and McKusick, V. A. (1953). *A.M.A. Arch. intern. Med.*, 92, 701.
 Shikhar, P. V. (1921). *Indian J. Med.*, 2, 326.
 Skipper, E., and Flint, F. I. (1952). *British Medical Journal*, 2, 9.
 Trias de Bes, L., Sanchez Lucas, J. G., and Ballesta Barcons, F. (1955). *Brit. Heart J.*, 17, 484.

INTERMITTENT HYDRARTHROSIS

BY

S. MATTINGLY, M.B., M.R.C.P., D.Phys.Med.

Late Senior Registrar, Department of Physical Medicine and Rheumatism, the Middlesex Hospital

"Of periodicity in disease, no more striking example can be quoted than the recurrent effusion into joints, with little or no febrile disturbance or local signs of inflammation, which goes by the name of intermittent hydrarthrosis."

Garrod (1910).

Intermittent hydrarthrosis is a condition of unknown aetiology characterized by transient joint effusions which recur at regular intervals; one or both knee-joints are usually affected, but occasionally other joints such as the hip, ankle, or elbow are involved (Ragan, 1949). The periodicity of the effusions was first noted in 1852 by C. H. Moore (1864), a surgeon at the Middlesex Hospital.

The onset is often sudden, the effusion lasting two to six days and recurring in cycles of 7 to 11 days, though extremes of 3 and 30 days have been reported. The duration of joint swelling and the length of each cycle

are remarkably constant in any one patient, although they may vary from time to time. If more than one joint is affected the effusions do not always coincide, but the cycle for each joint remains constant (Garrod, 1910; Bierring, 1921; Baker, 1929). When one joint ceases to swell another may become involved.

Both sexes are affected equally, usually between the ages of 20 and 50, although May and Robin (1947) and Ragan (1953) believe intermittent hydrarthrosis to be

FIG. 1.—Age at onset of 101 cases (52 male, 49 female) of intermittent hydrarthrosis reported in the literature.

most common in young women with onset at or after puberty. The age of onset and sex incidence of 101 cases reported in the literature does not support this view (Fig. 1).

Once established, intermittent hydrarthrosis may persist indefinitely, but is subject to spontaneous remissions and relapses which make assessment of treatment difficult. Although uncommon, it is probably not so rare as the literature would suggest, for fewer than 180 cases have been described. The following case reports may therefore be of interest.

Case 1

A housewife first noticed that her right knee was painful and swollen in April, 1950, at the age of 41. The swelling disappeared after a few days, only to reappear five days later, and thereafter the knee swelled at intervals of 9–13 days for a year. The swelling developed gradually, reaching its maximum in two to three days, and disappeared after five to six days. There was no history of previous injury, arthritis, or venereal disease, and the patient had never lived abroad. The effusions were not related to menstruation (Fig. 2).

In May, 1950, one month after the onset of the effusions, the patient developed an irritant rash over the lower trunk and thighs, thought to be true lichen planus; this persisted for over six months despite treatment with antihistamines, penicillin, vitamins, and local applications.

The patient remained in good health, and subsequent examinations revealed no abnormality except periodic effusions in the right knee-joint, accompanied at times by transient pitting oedema of the legs. There were no local signs of inflammation, and disability was not severe, though the patient felt unable to take up part-time domestic work.

The results of investigations, which included radiographs of the knee, haemoglobin estimation, white-cell count, erythrocyte sedimentation rate, and estimation of plasma proteins, were normal.

Treatment with pressure bandages, quadriceps exercises, antihistamines, autohaemotherapy, and penicillin was ineffective. In April, 1951, a course of gold injections was started, and in June, 1951, the patient reported that the knee had swollen only twice following chrysotherapy. Sodium aurothiomalate was given, by intramuscular injection, in weekly doses of 0.05 g. After the patient had received 0.75 g., 0.05 g. was given monthly to a total of 1.35 g. Injections were discontinued in August, 1953, as the patient went out to work and could not attend the clinic.

There has recently been a recurrence of her intermittent hydrarthrosis after a remission lasting five years.

Case 2

A housewife first noticed a painless swelling of the right knee in 1941, at the age of 27, when she was awakened in the early hours of the morning by an air raid and the knee "gave way." After two weeks the swelling disappeared, but six weeks later the left knee became swollen. For the next two years she was troubled by pain and swelling of alternate knees at intervals of three to four weeks. A remission lasting one year followed appendicectomy for acute appendicitis in December, 1943. Intermittent joint effusions, affecting alternate knees at intervals of two weeks, then recurred, each effusion lasting about four days. A further remission, lasting 10 months, followed venous thrombosis of the left leg with subsequent pulmonary infarction in January, 1951. The effusions recurred in November, 1951, again affecting alternate knees at two-weeks intervals, and antihistamines appeared to diminish pain and swelling without affecting the periodicity of the effusions.

The patient was admitted to hospital in May, 1952. Her general condition was good and examination of the knee-joints revealed no abnormality. Results of investigations, including haemoglobin estimation, white-cell count, erythrocyte sedimentation rate, serum uric acid estimation, and examination of catheter specimen of urine were normal; Wassermann reaction, gonococcal complement-fixation test, and brucella agglutination test were negative; radiographs of chest and hands were normal, and radiographs of the right knee showed early degenerative changes only. A small effusion, which developed in the right knee-joint, was aspirated; it was sterile and contained endothelial cells and lymphocytes only. Neither bed rest nor a four-day course of corticotrophin by injection influenced the periodicity of the effusions.

In June, 1952, the patient was treated as an out-patient with radiotherapy. Treatment was given to four areas, 10 by 8 cm., on the medial and lateral aspects of the knee-joints, the total skin dose to each area amounting to 1,200 r. The effusion recurred only once after radiotherapy.

FIG. 2.—Case 1. Chart showing periodic effusions in right knee-joint. Figures immediately above the line indicate length of cycles. Effusions not related to menstruation.

When last seen (November, 1955) there had been no recurrence of the effusions, although the patient's knees were sometimes stiff and painful if she was on her feet all day. No significant abnormality was found on examination and the erythrocyte sedimentation rate was normal.

Radiographs of the chest and sacro-iliac joints were normal, and there was no change in radiographs of the knees. The remission has lasted over four years.

The patient's aunt developed periodic swelling of the knees at the age of 63, following an attack of polyarthritis. No further details are known.

Case 3

A housewife first complained of pain in the knees in 1921, at the age of 19. She spent 18 months in bed with the joints immobilized in splints, and subsequently her tonsils were removed. In 1917 she had had diphtheria and in 1918 rheumatic fever.

She had no further trouble with her knees until 1929, when, three months after the birth of her second child, she developed a tubo-ovarian abscess and acute "rheumatism" in the legs. After seven months in bed at home and one month at a spa, she made a complete recovery and even took up ballroom dancing as a career. Two further relapses occurred in 1939 and 1946, but these lasted only a few weeks.

In the summer of 1948, at the age of 45, she developed intermittent hydrarthrosis. She noticed that painful swelling of the knees recurred every eight days. She felt very fit on the first morning of the eight-day cycle, but about 3 p.m. began to feel ill; fluid seemed to rush into the knee-joints, especially the right one, which swelled rapidly. She ran a fever, with temperatures up to 101° F. (38.3° C.), for a few days, and retired to bed. After four days she felt well, the swelling had subsided and she was free of pain for the next four days, when the cycle was repeated.

In 1949 she was admitted to hospital for investigation. On examination both knees were painful and swollen, flexion was limited to 90 degrees, and the quadriceps muscles were weak. The swelling was noted to subside and then recur at eight-day intervals, but the patient was not pyrexial during her stay in hospital. The haemoglobin, white-cell counts, and plasma uric acid were within normal limits, but the erythrocyte sedimentation rate was persistently raised (30–50 mm. in 1 hour, Wintrobe). Joint fluid was aspirated and found to be sterile and to contain only scanty polymorphs and lymphocytes; guinea-pig inoculation was negative. Radiographs of the chest and hands were normal. Radiographs of the knees showed some osteoporosis and early degenerative changes. Intradermal tests with foreign proteins suggested that the patient was sensitive to milk, fish, and eggs, but she was not treated with an exclusion diet. Bed rest, mepyramine maleate, 100 mg. t.i.d., and physiotherapy resulted in some symptomatic improvement, but a month later, after discharge from hospital, she complained of pain in many joints and was too ill to attend hospital as an out-patient.

Believing that no further treatment could help her, she accepted her disability. In 1951, two years later, the effusions suddenly ceased, but for a few months she experienced intermittent swelling of the eyelids at weekly intervals. The remission has now lasted for five years, although she says that she still feels unwell for a few hours every eight days.

When last seen (summer of 1956) she complained that while the legs were no longer swollen or painful they tended to give way in the street, and on several occasions she had fallen and injured herself. On examination she was found to have aortic incompetence. The knees were not swollen, but flexion was limited to 90 degrees and the quadriceps were weak and wasted. A full blood count was normal and no L.E. cells were found in the blood; the plasma uric acid was normal; the Wassermann reaction, and gonococcal complement-fixation, brucella agglutination, and Rose's differential agglutination tests were negative. Radiographs of the

chest, sacro-iliac joints, and hands were normal. Radiographs of the knees showed some osteoporosis and early degenerative changes only.

Discussion

The aetiology of intermittent hydrarthrosis remains obscure, and it may be a syndrome accompanying various joint disorders. The literature on the subject is confusing, because two groups of cases have been described: (1) "idiopathic," in which effusions in one or both knee-joints have recurred for 10–20 years without clinical or radiological evidence of permanent joint damage (Moore, 1864; Bierring, 1921; Miller and Lewin, 1924; May, Huet, and Barraud, 1937; Cook, 1937; Railliet, 1938; Weiner and Ghormley, 1956); and (2) "symptomatic," in which periodic effusions have been noted in patients already suffering from brucellosis (Baker, 1929), post-venereal arthritis (Garrod, 1910; Porter and Lonergan, 1932), ankylosing spondylitis (Porter and Lonergan, 1932), and rheumatoid arthritis (Garrod, 1910; Nielson, 1922; Steinberg, 1955).

Although intermittent hydrarthrosis was first thought to be due to malaria and was treated with quinine, there is little evidence that it is due to infection. Wallis (1955) has suggested that patients with "periodic diseases" may have chronic brucellosis. However, apart from one case described by Baker (1929), in which *Brucella melitensis* was cultured from joint fluid, blood, and urine, and a further doubtful case reported by Sharpe (1936), brucellosis has not been shown to be a cause of intermittent hydrarthrosis. Brucella agglutination tests are usually negative, as they were in the three cases here described. The joint fluid is usually sterile.

Cohen (1955) believes that all patients with idiopathic intermittent hydrarthrosis should be investigated as "potential arthritics of the rheumatoid type," and Ropes (1944) quotes the view of American physicians that it is a phase in the development of rheumatoid arthritis. Certainly intermittent hydrarthrosis is distinguished from palindromic rheumatism (Hench and Rosenberg, 1944) mainly by its periodicity and tendency to involve only the knee-joints, and cases diagnosed as palindromic rheumatism may, at a later date, develop joint damage and deformity (Kuhns, 1945).

Trauma (Krida, 1933; Railliet, 1938) and allergy (Lewin and Taub, 1936; Service, 1937; Pellet, 1946; Miller, 1949) have been incriminated. However, evidence of skin sensitivity to allergens is not proof that these are causative agents, and exclusion diets have proved disappointing in treatment.

Angioneurotic oedema is occasionally periodic, and Fridenberg (1888) suggested that it might be related to intermittent hydrarthrosis. Garrod (1910) noted the relationship between transient joint lesions and erythematous skin lesions in Henoch's purpura, erythema nodosum, and rheumatic fever. Recurrent circumscribed oedema of lips or eyelids accompanied intermittent hydrarthrosis in one of his patients with chronic rheumatoid arthritis, and he quotes the case of a woman of 60, with a seven-year history of rheumatoid arthritis, who was subject to periodic subcutaneous swellings without effusion into joints. Berger (1939) described the case of a woman of 22, in whom joint swellings promptly disappeared on an exclusion diet, to be replaced by angioneurotic oedema. It is interesting that Case 3, above, had intermittent swelling of the eyelids after joint swellings had ceased.

Reimann (1948) related intermittent hydrarthrosis to periodic fevers, neutropenia, and myasthenia, and noted occasional periodicity in migraine, epilepsy, and psychoses. These episodic diseases did not appear to coincide with menstruation, but were affected by pregnancy. He suggested that they might represent exaggerated forms of similar but symptomless rhythms in normal people. Although intermittent hydrarthrosis may begin with the menarche and cease during periods of amenorrhoea (Hiller, 1952), as well as during pregnancy and lactation, it has not yet been proved an endocrine disorder. Reports of successful treatment with

thyroid extracts (Pellet, 1946) and testosterone (Lièvre, 1947; Hiller 1952) are unconvincing.

There is no evidence that the condition is inherited, and a familial incidence has been reported only once before.

There have been few reports on pathological changes in the synovia. Ghormley and Deacon (1936) found slight thickening of the lining cells only. Krida (1933) performed a synovectomy during an acute attack; the synovial membrane was pale, pink, and thickened, and microscopy revealed hyperaemia, round-cell infiltration, and granulation tissue in the enlarged villi and subsynovial tissues. Similar findings were reported by Weiner and Ghormley (1956). Ragan (1953) also found changes indistinguishable from rheumatoid arthritis, although his patient had a normal erythrocyte sedimentation rate and a complete remission without significant joint changes. With one exception (Baker, 1929) the joint fluid has been sterile, but it may contain several thousand cells per cubic millimetre, with over 50% polymorphs (Collins, 1936). The hyaluronic acid in the synovial fluid of the cases seen by Ragan (1953) was normal, in contradistinction to the changes which occur in rheumatoid arthritis. In "idiopathic" intermittent hydrarthrosis the sedimentation rate, haemoglobin level, and white-cell count are normal; the Wassermann reaction and gonococcal complement-fixation and brucella agglutination tests are negative, as are blood cultures. When intermittent hydrarthrosis develops in a patient with a chronic rheumatoid type of arthritis the results of investigation are those usually found in that disease.

Although the characteristic feature of the condition is the periodicity of the effusions, the cycle of joint swellings is perhaps not so constant as May, Huet, and Barraud (1937) believe when they state that "at the beginning of the year the patients can mark on their calendars in advance the dates on which they will be indisposed." Garrod (1910) pointed out that when the effusions are charted for long periods the attacks are not so regular as they appear to be if only a few effusions are observed, and this was so in Case 1 (Fig. 2). In the idiopathic cases, signs of local inflammation such as heat, tenderness, and muscle spasm are lacking and there is no constitutional disturbance. Transient oedema of the legs, as in Case 1, was also noted by May and Robin (1947).

Spontaneous remissions frequently occur and may last 5-10 years, making assessment of treatment difficult. Relapses, for no apparent reason, are equally common and the condition may persist indefinitely. The great majority of cases reported in the literature have been followed up for less than three years. While some of the patients with "idiopathic" intermittent hydrarthrosis undoubtedly develop rheumatoid arthritis, there is no evidence that this is inevitable or a frequent occurrence. Cohen (1955) has even seen a patient whose attacks recurred for over 30 years without developing rheumatoid arthritis, and Ragan (1953) states the prognosis of these cases to be good with regard to eventual crippling, although attacks may continue without remission. None of the 47 patients described by Weiner and Ghormley (1956) developed rheumatoid arthritis.

Many of the early cases of intermittent hydrarthrosis were treated with quinine, for periodicity was thought to be synonymous with malaria. It rarely proved beneficial, and other treatments equally unsuccessful included tonsillectomy, autohaemotherapy, histamine desensitization, and the administration of arsenic, mercury, ovarian extracts, stilboestrol, chorionic gonadotrophin, antihistamines, and cinchophen; injections of adrenaline, calcium gluconate, and "para-thormone" proved useless. Bed rest, ice packs, mud baths, splints, and exercises have been ineffective.

Exclusion diets, following skin tests for allergens, were credited with remissions by Berger (1939) and Miller (1949), but the latter failed to help a further case, and Epstein and Edeiken (1940) were also disappointed. Weismann-Netter (1929, 1937) claimed success with ergotamine tartrate by injection and by mouth respectively. The period of follow-up in both cases was short, and the first patient had a severe

reaction to treatment. Perhaps more convincing is the report of a case treated by Cook (1937): a man of 43, with a 15-year history of intermittent hydrarthrosis, remained in remission for three years following a prolonged course of ergotamine tartrate, 1 mg. daily by mouth. Ragan (1949) and Railliet (1938) were less successful, and most authors stress the dangers of long-term administration of this drug.

Temporary remissions have been produced by intravenous T.A.B. vaccine, but Cohen (1955) emphasizes the danger of a severe reaction; a generalized arthritis may result.

Gold injections have not previously been advocated in the treatment of intermittent hydrarthrosis, which is surprising, as they have been found to benefit patients with palindromic rheumatism (Boland and Headley, 1949) and patients with an episodic or palindromic type of rheumatoid arthritis. The remission reported in Case 1 lasted more than five years, and this result is encouraging enough to suggest a further trial of chrysotherapy.

There are few reports on the use of steroids in treatment. Steinberg (1955) found oral cortisone, 25 mg. t.i.d., was ineffective, and intra-articular hydrocortisone, whilst having a temporary effect, may aggravate the condition. A short course of corticotrophin by injection failed to prevent the onset of an attack in Case 2.

Few patients have received chemotherapy. Case 1 had two courses of intramuscular "distaquine" (300,000 units daily for five days, and 600,000 units daily for 20 days) for minor skin sepsis complicating lichen planus, without affecting the effusions. Intra-articular injection of antibiotics has not been reported and might be more effective than systemic administration.

Temporary remissions have resulted from radiotherapy (May, Huet, and Barraud, 1937; Epstein and Edeiken, 1940; Ragan, 1953), but patients were followed up for less than two years, and Ragan (1949), Lièvre (1947), and Railliet (1938) found radiotherapy ineffective. Case 2 has now had a remission lasting four years following radiotherapy, and, whilst it is too early to decide that the remission is complete, the patient remains very grateful.

Synovectomy has been recommended, but Krida (1933) followed up his two cases for only a year, and Key (1940) gives no details of cases so treated. Ragan (1949) found that, while synovectomy prevented recurrence of effusions in a knee-joint for over five years, the operation was followed within a few months by recurrent effusions in both elbow and hip-joints. Weiner and Ghormley (1956) found synovectomy was effective in a few patients, but noted recurrence of the effusions after an interval of one to two years in several joints.

Summary and Conclusions

Intermittent hydrarthrosis is characterized by periodic joint effusions, usually affecting one or both knee-joints.

The cause is not known. Some patients develop rheumatoid arthritis.

Spontaneous remissions and relapses are common and make assessment of prognosis and treatment difficult.

Three cases are described; in two, remissions were apparently induced by chrysotherapy and radiotherapy.

Intermittent hydrarthrosis may be an unusual variant of rheumatoid arthritis, but permanent joint damage is not inevitable. It could, however, be a syndrome accompanying various joint disorders, the periodicity of the effusions being related to normal but exaggerated cyclical changes in the tissues of the patient.

Treatment is, of necessity, empirical and usually ineffective; radiotherapy, chrysotherapy, and synovectomy may induce at least a temporary remission.

I am indebted to Dr. A. C. Boyle, of the Middlesex Hospital, for his help with this paper and permission to publish details of Cases 2 and 3, and to Dr. H. A. Burt, of University College Hospital, for permission to report Case 1.

REFERENCES

- Baker, B. M. (1929). *Arch. intern. Med.*, 44, 128.
 Berger, H. (1939). *J. Amer. med. Ass.*, 112, 2402.
 Biering, W. L. (1921). *Ibid.*, 77, 785.
 Boland, E. W., and Headley, N. E. (1949). *Ann. rheum. Dis.*, 8, 64.
 Cohen, H. (1955). In Copeman's *Textbook of Rheumatic Diseases*, 2nd ed., p. 276. Livingstone, Edinburgh.
 Collins, D. H. (1936). *J. Path. Bact.*, 42, 113.
 Cook, C. K. (1937). *Med. Rec. (N.Y.)*, 145, 300.
 Epstein, H. J., and Ekelken, L. (1940). *Radiology*, 35, 360.
 Fridenberg, A. H. (1888). *Med. Rec. (N.Y.)*, 33, 657.
 Garrod, A. E. (1910). *Quart. J. Med.*, 3, 207.
 Ghormley, R. K., and Deacon, A. E. (1936). *Amer. J. Roentgenol.*, 35, 740.
 Hench, P. S. and Rosenberg, E. F. (1944). *Arch. intern. Med.*, 73, 293.
 Hiller, R. I. (1952). *Amer. J. Obstet. Gynec.*, 64, 209.
 Key, J. A. (1949). *J. Amer. med. Ass.*, 115, 2209.
 Krida, A. (1933). *J. Bone Jt Surg.*, 15, 449.
 Kuhns, J. G. (1945). *Arch. Surg. (Chicago)*, 51, 289.
 Lewin, P., and Taub, S. J. (1936). *J. Amer. med. Ass.*, 106, 2144.
 Lièvre, J. A. (1947). *Bull. Soc. méd. Hôp. Paris*, 63, 183.
 May, E., Huet, J. A., and Barraud, M. (1937). *Ibid.*, 53, 106.
 — and Robin, J. (1947). *Presse méd.*, 55, 129.
 Miller, J. (1949). *Ann. Allergy*, 7, 497.
 Miller, J. L., and Lewin, P. (1924). *J. Amer. med. Ass.*, 82, 1177.
 Moore, C. H. (1864). *Lancet*, 1, 485.
 Nielson, A. L. (1922). *J. Amer. med. Ass.*, 78, 507.
 Pellet, C. (1946). *Rev. Rhum.*, 13, 338.
 Porter, J. L., and Loneragan, R. C. (1932). *J. Bone Jt Surg.*, 14, 631.
 Ragan, C. (1949). In *Comroe's Arthritis and Allied Conditions*, 4th ed. Kimpton, London.
 — (1953). *Ibid.*, 5th ed.
 Railliet, G. (1938). *Bull. Soc. méd. Hôp. Paris*, 54, 1579.
 Reimann, H. A. (1948). *J. Amer. med. Ass.*, 136, 239.
 Ropes, M. W. (1944). *Bull. New Engl. med. Cent.*, 6, 54.
 Service, W. C. (1937). *Amer. J. Surg.*, 37, 121.
 Sharpe, J. C. (1936). *Ann. intern. Med.*, 9, 1431.
 Steinberg, V. L. (1955). *Ann. phys. Med.*, 2, 255.
 Wallis, H. R. E. (1955). *Lancet*, 1, 1331.
 Weiner, A. D., and Ghormley, R. K. (1956). *J. Bone Jt Surg.*, 38, 1039.
 Weissmann-Netter, R. (1929). *Bull. Soc. méd. Hôp. Paris*, 45, 909.
 — (1937). *Ibid.*, 53, 185.

ACUTE SCHÖNLEIN-HENOCH PURPURA TREATED WITH PREDNISOLONE

REPORT OF A CASE

BY

ALAN S. COHEN, A.B., M.D.

Lately Registrar in Medicine, Dundee Royal Infirmary,
Dundee

Schönlein-Henoch purpura is a disease associated with an increased permeability of the capillary endothelium permitting the passage of blood cells and plasma (Wintrobe, 1946). The specific aetiology is not clear (Davis, 1948), although evidence points to a hypersensitivity to an antigen which may be bacterial or non-bacterial (Gairdner, 1948; Jensen, 1955). Prognosis is dependent upon whether or not severe renal damage is caused (Ackroyd, 1953). Until recently it was generally believed that no medication had a favourable effect on the natural course of the disease (Davis, 1948; Gairdner, 1948). When the adrenal and pituitary steroids were found to exert favourable responses in various allergic disorders, several investigators studied their effect on Schönlein-Henoch purpura. The results have been variable, but a symptomatic response has been observed in at least half of the reported series (Stefanini *et al.*, 1950; Levinson *et al.*, 1951; Adamson *et al.*, 1953; Leese, 1955). It was therefore of interest to observe the effect on the course of the disease of prednisolone. Stefanini and Martino (1956) stated that this drug had caused striking remissions in two cases of the acute variety of Schönlein-Henoch purpura.

The following is a report of a typical case of this disease complicated by severe and progressive renal involvement. The downhill course was apparently unaffected by cortisone, but was dramatically reversed by prednisolone.

Case Report

A 10-year-old schoolgirl was transferred to the Dundee Royal Infirmary because of purpuric eruption and rectal bleeding. She apparently had been in excellent health until three and a half weeks previously, at which time she had an abrasion of her knee requiring no specific treatment. The following day she had had an uneventful dental extraction. Four days later she complained of abdominal discomfort and vomiting. Her family doctor found she had a fever and gave her 500,000 units of penicillin. Her symptoms continued, and, as she was also suspected of having tonsillitis, she was admitted to a fever hospital one week after the onset of her illness. At that time physical examination was negative except for enlarged tonsils. Her white count, sedimentation rate, haemoglobin, urinalysis, and chest x-ray films were all within normal limits.

Within several days of admission, however, scattered haemorrhagic spots appeared over her arms. These soon spread to involve her back, buttocks (see Fig. 1), and lower extremities. Two days after the appearance of the rash, gross blood was observed in her stools. She also complained of stiffness in her right knee and both shoulders and elbows, though no redness, swelling, or tenderness was noted in these areas. The following day she had pain in the joints

FIG. 1.—Haemorrhagic eruption on buttocks of a 10-year-old girl with Schönlein-Henoch purpura; second week of illness.

of the left hand and a small effusion was found over the dorsum. New crops of haemorrhagic spots were constantly appearing. Albuminuria occurred, and one week after the appearance of the bloody stools fresh blood was found in her urine. A diagnosis of anaphylactoid, or Schönlein-Henoch, purpura was therefore made, and the patient was transferred to the Royal Infirmary approximately three and a half weeks after the initial onset of her symptoms.

At this time she was an acutely ill, pale girl complaining of colicky abdominal pain, which came in spasms that lasted from five to ten minutes. A haemorrhagic eruption was present over buttocks, abdomen, and extremities; the face was clear. The rash varied from fresh spots to faded areas. Examination of heart and lungs was negative. Blood pressure was 130/80. Abdominal examination showed no enlargement of liver or spleen, and though some tenderness was present the abdomen was soft throughout. Neurological examination was negative. Joints were normal. Laboratory studies showed: haemoglobin, 8.8 g. per 100 ml.; white blood cell count, 16,000 per c.mm.; E.S.R., 35 mm. in 1 hour; blood urea, 20 mg. per 100 ml.; cholesterol, 120 mg. per 100 ml.; serum bilirubin 4 mg. per 100 ml.; while the serum sodium, potassium, chloride, and carbon-dioxide-combining power were all within normal limits. The platelet count was 851,000 per c.mm. The antistreptolysin titre (drawn four weeks after admission) was within normal limits. X-ray films of chest and abdomen and electrocardiograms were