THE SCHOENLEIN-HENOCH SYNDROME IN CHILDHOOD WITH PARTICULAR REFERENCE TO THE OCCURRENCE OF NEPHRITIS

BY

M. G. PHILPOTT

From the Department of Child Health of the University of Sheffield

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The full syndrome of a purpuric skin eruption, intestinal bleeding and swollen joints was first described by Henoch (1874). Since that date there have been numerous papers about the syndrome. Gairdner (1948) reviewed the literature and grouped the condition with acute rheumatism, acute nephritis and polyarteritis nodosa. The name 'Schoenlein-Henoch Syndrome' used by Gairdner is adhered to in the present communication.

Attention to the association of the syndrome with nephritis was drawn by Henoch (1899), and this was amplified by other authors (Macalister, 1906; Osler, 1914). Nephritis is now recognized as being the most serious complication of the disease. Gairdner (1948) found evidence of renal involvement in all but one of his 12 cases. Haematuria was reported in four of 44 cases described by Davis (1948) and in 10 of 20 cases described by Balf (1951). In neither of these papers was it made clear in all cases whether the haematuria was macroscopic only or whether microscopic haematuria was also included, and neither author mentioned whether complete and repeated urine analyses were made in their cases. The 44 cases reported by Davis included adults as well as children, the ages ranging from 4 to 71 years. Unfortunately he did not state the proportion of children in his series.

No paper has been found which sets out specifically to determine the incidence of nephritis in a large series of cases. This paper describes the relapse rate in 40 cases of the Schoenlein-Henoch syndrome in children, with the incidence and progress of complicating nephritis.

Material

Only cases in which there was no doubt as to diagnosis, each exhibiting at least the typical rash of maculo-papules and petechiae and either abdominal or joint symptoms and having normal platelet counts, have been admitted to this study. All the patients attended either the Children's Hospital, Sheffield, or the paediatric department of the City General Hospital, Sheffield, during the

five-year period of January, 1947, to December, 1951.

The urine was examined microscopically and chemically regularly for at least two months after the start of the disease and one month after clinical recovery. Twenty-seven cases were followed-up for periods ranging from six months to five years after the onset of the disease. Of the remaining patients, in eight the disease was of too recent onset to include them in the group followed-up, and five could not be traced. In children whose urine contained more than 10 mg. % of protein and red blood cells and casts in the centrifuged deposit, 12-hour Addis counts were made, using the criteria of Giles (1947) for normality.

Results

Occurrence of Nephritis. Forty cases were examined in the acute stage of the disease. Of these, 19 (47.5%) had evidence of renal involvement. There were protein and casts in the urine of all; in four there was macroscopic haematuria while in the remainder the haematuria was microscopic only. In five cases the abnormalities were found on admission and in none did they occur after six weeks from the start of the disease.

In three of the four who had macroscopic haematuria this came on suddenly one to five weeks after the beginning of the disease. In one case nephritis had been present for 11 months before the onset of the Schoenlein-Henoch syndrome, and the nephritis had been preceded by acute rheumatism. Macroscopic haematuria recurred with the rash.

Prognosis of Nephritis. Of the 19 children who developed evidence of renal involvement, 11 have been followed up for a year or more from the time that urinary abnormalities were first noted. Seven of these have shown complete clearing of the nephritis, as evidenced by a normal Addis count and a normal blood pressure. The other four still have evidence of active nephritis, including the one case in which the nephritis preceded the Schoenlein-Henoch syndrome. Sixteen cases in which no renal involvement was found at the time of the active disease have been followed up for six months or

more. None of these showed any urinary abnormalities at any time.

Relapses and Recurrences. In attempting to ascertain what proportion of cases suffer relapses and recurrences, a return of the skin eruption, sometimes associated with abdominal or joint symptoms, occurring within three months of clinical recovery is termed a relapse, and occurring later than three months a recurrence. Of 27 cases followed up for six months or more none suffered recurrences. Of the whole 40 cases, 14 (35%) suffered relapses. The skin eruption relapsed in some cases as often as six times. In one case such episodes occurred over a period of seven months. Abdominal and joint symptoms only occasionally relapsed and then only once or twice. Of the 14 cases relapsing, 12 developed nephritis, usually during the first or second attack, while of 26 cases without relapses only seven developed nephritis.

Aetiological Factors. Unfortunately the bacteriology of the throat was studied in only a few cases. However, a history of sore throat occurring in under four weeks before the onset of symptoms was recorded in 20 cases, and in two further cases without a history of sore throat a moderate growth of beta-haemolytic streptococci was obtained from throat swabs taken on admission. Thus at least 22 (55%) cases suffered throat infections before the onset of the disease.

In no case was there definite evidence of allergy to food. In one case there was a sore throat and beta-haemolytic streptococci were grown from the throat swab on admission. Three relapses occurred, each after the ingestion of ice cream. Further relapses occurred, however, without the ingestion of ice cream, and at a later date further consumption of this food did not cause relapses. In two cases without histories of sore throats food allergens may have been responsible, canned meat in one and eggs in the other, but no relapses occurred when these foods were given again.

Age and Sex Incidence. The youngest patient was only 8 months old, while the oldest was 10½ years. Four patients were under 2 years of age. The average age was 5.2 years. Of the 40 patients, 25 were boys and 15 girls.

Discussion

In an analysis of the results of this survey the high incidence of nephritis is worthy of note (19 out of 40 cases or 47.5%), especially in those who had relapses (12 out of 14 cases). The number is too small to permit the assessment of the prognosis of nephritis in this disease. Out of 11 cases in the present series four still had active nephritis after

one year. Macalister (1906), Osler (1914) and Gairdner (1948) have all drawn attention to the severity and poor prognosis of this complication.

Although 14 of the 40 children relapsed, none of 27 followed-up for more than six months suffered recurrences. Twenty-two of the 44 cases reported by Davis (1948), many of whom were adults, suffered relapses and recurrences. Recurrences after three months of freedom from symptoms appear to be rare in children.

Few authentic cases have been reported in children under the age of 1 year and the disease is rare under the age of 2 years. The child aged 8 months must be regarded as a rarity. The only case under this age found in the literature was an infant of 7 months reported by Scolpini, Garcia Rodriguez and Angelillo (1948), but the diagnosis was uncertain as the infant had definite thrombocytopenia, and the history was not completely typical of the Schoenlein-Henoch syndrome.

Gairdner (1948), in reviewing the literature, found that there was a higher incidence in boys than in girls but Davis (1948) reported a heavy preponderance of females in his mixed series of adults and children. The present findings correspond with those of Gairdner.

The significance of a history of throat infection occurring in over 50% of the cases is difficult to determine in the absence of bacteriological investigations. It is at least probable that there is a fairly high incidence of throat infections in cases of this disease.

Summary

Forty cases of the Schoenlein-Henoch syndrome in children have been analysed. Nineteen (47.5%) developed nephritis. Fourteen (35%) suffered relapses and 12 of these developed nephritis. Of 11 patients developing nephritis, this is still active in four after one year. Only four cases were encountered under the age of 2 years, including one infant of 8 months. Male cases were commoner than female in a ratio of 5:3. Throat infections preceded the disease in at least 55% of the cases.

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* Standard error = $\pm 16\%$.