SUMMARY

In a retrospective survey of renal amyloidosis in a large general hospital, only 7 cases were found. Patients generally presented with nephrotic syndrome and symptoms of fluid overload; hypertension on presentation was unusual. Renal failure was present in 5 out of 7 patients, and uraemia in 3. The disease was secondary in 5 patients and primary in 2, and the prognosis was uniformly bad.


Amyloidosis, a disease characterized by the extracellular tissue deposition of an amorphous hyaline material, has been well recognized since its description by Rokitansky in 1842. The material has a homogeneous eosinophilic appearance on histological sections, it has an affinity for Congo red and displays a green birefringence when examined in polarized light, and a fibrillar structure is demonstrable by electron microscopy.

No entirely satisfactory classification of amyloidosis exists, but the disease occurs in several distinguishable forms, namely (i) primary amyloidosis, where no previous or coexisting disease is demonstrable; (ii) secondary amyloidosis, after a number of chronic supplicative or inflammatory diseases; (iii) amyloidosis associated with myelomatosis and other tumours; and (iv) heredofamilial amyloidosis, especially associated with familial Mediterranean fever.

Secondary amyloidosis has been described in association with a large number of conditions, including tuberculosis, leprosy, syphilis, bilharziasis, bronchiectasis, empyema, lung abscess, osteomyelitis, paraplegia, ulcerative colitis, regional enteritis and rheumatoid arthritis. In all varieties of amyloidosis, the most important site of involvement is the kidney, although liver, heart, spleen,
blood vessels, gastro-intestinal tract, skin and nerves may also be involved.

Some of the conditions known to predispose to secondary amyloidosis are rare in Blacks (e.g. rheumatoid arthritis, regional enteritis, ulcerative colitis), while others (e.g. tuberculosis and leprosy) are common.

It is the purpose of the present study to describe those patients with documented amyloidosis encountered over a 3-year period in a large general hospital for Blacks in Johannesburg, and to compare the clinical features with those described in the literature, with particular reference to some of the more controversial aspects of the disease.

SUBJECTS AND METHODS

The case records of patients with renal amyloidosis, diagnosed by percutaneous biopsy, seen over a 3-year period at Baragwanath Hospital, were reviewed. In all patients the diagnosis was unequivocally established by the demonstration of green birefringence under polarized light, in tissue stained with Congo red. All pertinent clinical and laboratory data were extracted and analysed.

RESULTS

Over the 3-year period reviewed, 7 patients with renal amyloidosis were encountered. Their clinical data are shown in Table I. There were 4 males and 3 females with ages ranging from 23 to 77 years; all presented with symptoms caused by fluid overload; 1 had pulmonary oedema, 4 had combined pulmonary and peripheral oedema, and 2 had peripheral oedema only. Hepatomegaly was found in 5 of the 7 patients. Only 1 patient was hypertensive.

The amyloidosis was associated with, and presumably secondary to, bronchiectasis in 1 patient, leprosy in 2 patients, tuberculosis in 1 patient, and bilharziasis in 1 patient. One patient had massive lymphoedema of both legs, but no primary disease was demonstrable, even at autopsy, while in another patient no other disease was detected clinically. These 2 patients were classified as having primary amyloidosis.

The haematological and biochemical features are shown in Table II. Two patients had normal or borderline renal function, while the other 5 had renal failure of greater or lesser severity.

All 7 patients had hypo-albuminaemia (<2,5 g/dl), and in 6 of the 7 in whom it was documented, there was heavy proteinuria (>3,5 g/24 hours). All patients displayed the features of the nephrotic syndrome despite the marked reduction in renal function in 3. Four of 6 patients had hypergammaglobulinaemia, but none had abnormal proteins on electrophoresis (Table III).

In 6 of the 7 patients the renal biopsy was performed as part of the evaluation of the nephrotic syndrome. The seventh patient presented with uraemia and large kidneys, and renal biopsy was performed for this reason. In 2 of the 7 patients, hepatic involvement was also demonstrated histologically. Rectal biopsy was performed in 4 patients, and amyloid was present in only 2 (Table IV).
There was no long-term follow-up in 3 of the 7 patents. Of the other 4, 2 had end-stage renal failure and died after refusing haemodialysis, 1 died suddenly after 9 months of regular haemodialysis, while the fourth died after a protracted illness. Autopsy was performed only on the patient with massive lymphoedema. Amyloid was demonstrated histologically in kidney, liver, spleen, and in the coronary and systemic arteries. There was no clear cause for the massive lymphoedema, nor was a primary disease demonstrated at autopsy (Table IV).

### DISCUSSION

By all accounts, amyloidosis is a rare disease. It was encountered in 21 instances in a series of 4000 autopsies performed at the Massachusetts General Hospital. The majority of these cases were secondary to rheumatoid arthritis or multiple myeloma. In another series of 20 000 autopsies done at Johns Hopkins Hospital, there were 79 cases of amyloidosis.

The diagnosis of amyloidosis depends on the finding of deposits of amyloid material in appropriately stained tissue specimens. The Congo red (Paunz) test has fallen into disuse. Less invasive biopsy procedures are gingival biopsy, rectal biopsy and needle biopsy of subcutaneous fat, while demonstration of amyloid fibrils in the urinary sediment may be diagnostic. Renal biopsy is positive more often than rectal biopsy but is difficult and hazardous, as is liver biopsy. Electron microscopy may occasionally reveal amyloid fibrils when other tests have been negative.

Renal involvement in secondary amyloidosis may be viewed in one of two ways. In the context of amyloid disease in general, renal involvement has been reported to occur in between 83% and 100% of cases. On the other hand, in the context of renal disease generally, for example in biopsy specimens of kidney tissue, Heptinstall and Joekes found amyloid disease in 3% of a large series of renal biopsies, while Walker et al. only encountered 7 cases in a series of about 300 biopsies. The 8 cases reported here occurred in a series of about 150 biopsies performed over the 3-year period reviewed. When viewed against the prevalence of tuberculosis in South Africa, amyloidosis is indeed a rare disease.

Renal involvement in amyloidosis varies with the underlying condition, occurring in 75% of cases of secondary amyloidosis, one-third of cases of multiple myeloma and in one-quarter of cases of primary amyloidosis. Dunhill, however, reports that the incidence of renal involvement is the same in primary and secondary amyloidosis. Of the 79 cases in the series of Walker et al., 46 were associated with chronic suppuration, 5 with myeloma, and 28 were cases of primary amyloidosis. Of the 79 cases, renal involvement was demonstrated in 16 and was especially associated with chronic suppuration. On the other hand, in a large series of 236 cases, 193 (82%) were either primary (56%) or myeloma-associated (26%).

The symptomatology of renal involvement in amyloidosis is variable. It may be completely asymptomatic, proteinuria being discovered incidentally, while more advanced involvement may be manifested by the nephrotic syndrome or by progressive uraemia. All 8 patients in the present series presented with fluid retention (peripheral and/or pulmonary oedema) suggesting the presence of more advanced disease than that reported in other series.

Renal failure is reported to be an uncommon or late manifestation occurring in 50% of cases in one series. Six of the 7 patients in the present series had impaired renal function; 3 had end-stage disease at presentation and the fourth progressed rapidly to end-stage disease in little over a year.

Another controversial point in amyloidosis is the frequency of hypertension. Although generally regarded as infrequent, the reported incidence has varied from as low as zero to as high as 35 - 50%. and is said to be more frequent in the presence of advanced renal disease and azotaemia. Only 1 of the 8 patients in the present series had hypertension on admission. He, however, presented with pulmonary oedema and as soon as his fluid overload was improved by dialysis, his blood pressure dropped to normal levels and remained so throughout the 9 months that he was on regular haemodialysis.

What is generally agreed upon in the literature is that, with few exceptions, amyloidosis is a progressive disease and is almost invariably fatal, and that renal involvement is associated with the poorest prognosis. Of 42 patients in one series, 43% died of uraemia. In a study of 48 patients followed up for as long as 14 years, the mean survival time was 44 months; only 6 patients lived for 5 years or longer, and renal failure or its complications was the usual cause of death. Even with the advent of haemodialysis, the life expectancy of patients with renal failure due to amyloidosis is not very much prolonged. The death of all 4 patients with severe renal failure in the present series confirms this high mortality. The 1 patient who was on regular haemodialysis also died after only 9 months of treatment.

| TABLE IV. HISTOLOGICAL SITES OF AMYLOID DEPOSITION AND OUTCOME OF PATIENTS |
|----------------|----------------|----------------|
| Case | Kidney | Liver | Rectum | Comment |
| 1   | +     | +     | -      | No follow-up |
| 2   | +     | -     | -      | Refused haemodialysis. Died |
| 3   | +     | +     | ?      | Refused haemodialysis. Died. Also found in spleen, blood vessels postmortem |
| 4   | +     | -     | 0      | No follow-up |
| 5   | +     | -     | +      | No follow-up |
| 6   | +     | +     | -      | Died after 9 months of haemodialysis |
| 7   | +     | +     | 0      | Reported elsewhere. Died |

No follow-up
Refused haemodialysis. Died
Refused haemodialysis. Died. Also found in spleen, blood vessels postmortem
No follow-up
No follow-up
Died after 9 months of haemodialysis
Reported elsewhere. Died
Breast Feeding in Cape Town

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SUMMARY

An interview survey of infant feeding practices was conducted among the mothers of 126 infants, aged 3 - 4 1/2 months, attending local authority clinics in two socio-economic residential areas of Cape Town. It was found that while at 1 month only 19% of the infants were wholly bottle-fed, at the age of 3 - 4 1/2 months this proportion was 60%, and only 14% were fully breast-fed. The factors found to be significantly associated with the cessation of breast feeding were young maternal age and maternal employment.

A downward trend in the prevalence of breast feeding continues — despite the efforts of health services. Recommendations to promote breast feeding are made.


Evidence continues to accumulate as to the benefits of breast feeding young infants. The advantages include the prevention of infection, allergy, rickets and iron deficiency, and the enhancement of mother-infant bonding. The economic advantages remain greater than ever. Recently, vigorous efforts to promote breast feeding have been made by health workers, particularly those engaged in obstetrics, neonatal care and child welfare services. This study was undertaken, firstly, to assess the impact of such efforts on the prevalence of breast feeding in an area of Cape Town; secondly, to establish factors associated with failure of breast feeding; and thirdly, to make recommendations concerning the promotion of breast feeding in the community.

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REFERENCES


SUBJECTS AND METHODS

The subjects were infants attending two local authority child welfare clinics in Heideveld and Manenberg — residential areas on the Cape Flats. The community served by both clinics is largely working class, occupying local authority housing schemes, in which a large degree of overcrowding exists. Infant mortality for this community in 1977 was 25.9 per 1000 live births. The main causes of infant death were prematurity, pneumonia and gastro-enteritis, which accounted for the deaths of 2 out of 3 infants under 1 year of age. A very high proportion of women are delivered in institutions, but most are discharged within 48 hours of delivery.

As part of a study on infant nutrition, all attendances at each clinic were monitored during two consecutive 2-week periods in March 1978. The mother of every infant in the age group 3 - 4 1/2 months was interviewed by a social worker (R. de W.). In addition to basic personal data, mothers were asked about employment, current contraceptive practice, place of delivery, method of feeding, and the duration of breast feeding. Data were processed and analysed by the Data Processing Unit of the Institute of Child Health, University of Cape Town.

Feeding categories were considered as follows: (i) wholly breast-fed; (ii) breast- and bottle-fed; and (iii) wholly bottle-fed.

RESULTS

Mothers

Maternal age groups are shown in Fig. 1. The modal age group was 21 - 25 years. Overall, 34% of mothers were unmarried. This figure is slightly lower than that for the general population. At the time of interview, 80% of the mothers were not at work; 2% were in part-time work, and 18% were in full-time work. Significantly more