Hospital Topics

Acute admissions of patients with sickle cell disease who live in Britain

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Abstract

All acute admissions of patients with sickle cell disease who lived in the London borough of Brent and attended this hospital were analysed for a period of one year. Sixty three of the 211 patients who were followed up by the haematology department required 161 acute admissions during the year. Most admissions (126) were for the 42 patients with homozygous sickle cell disease; 147 (91%) were for vaso-occlusive episodes, 142 of which were for painful crises, three for cerebrovascular accidents, and two for renal papillary necrosis. Preschool children with sickle cell disease were admitted predominantly with limb pain, whereas in schoolchildren and adults the incidence of trunk pain was higher. Twenty four of the 93 episodes of trunk pain culminated in an episode of severe visceral sequestration usually affecting the lungs, the liver, or the mesenteric circulation. Two patients died: an 18 month old baby with an acute splenic sequestration crisis and a 19 year old man with a severe girdle syndrome (sickling in the mesenteric circulation, liver, and lungs). Infective episodes were rare (11 episodes) but severe: one haemophilus meningitis, two salmonella infections, and three aplastic crises due to parvovirus infections. The average duration of the hospital stay was 7.4 days per admission.

It is concluded that because sickle cell disease causes appreciable morbidity in older children, adolescents, and adults a systematic approach to management is needed to deal with acute episodes such as sequestration syndromes.

Introduction

The number of patients with sickle cell disease can be predicted accurately if the number of people of Caribbean and African extraction in the population is known. The prevalence of sickle cell disease is 5.6 per 1000 births in Jamaica¹ and between 10 and 20 per 1000 births in different regions of West Africa.² From the 1981 census it can be calculated that at least 600 000 people of Caribbean extraction and 100 000 of African extraction were living in Britain at that time.³ If we assume that five per 1000 of the West Indians and 10 per 1000 of the West Africans have sickle cell disease the expected number of patients in 1981 was 4000 (3000 West Indians and 1000 West Africans).

This is in good agreement with the 1979 postal survey of sickle cell disease in England and Wales⁴: 1367 cases were identified and

the authors believed that the true number was at least twice as large (only haematologists were approached, only 60% responded, etc). From the Brent⁵ and Camberwell⁶ data on newborn screening for sickle cell disease it can be predicted that between 100 and 150 babies with the disease are born in Britain every year. If the lower estimate of 100 babies per year is taken the number of patients with the disease must have increased by 100 every year since 1981, and there are now at least 4500 patients with sickle cell disease in Britain. This number does not take into account new patients who immigrate or visit Britain, or the mortality. Nevertheless, 4500 appears to be a reasonable estimate. At present sickle cell disease is the most common haemoglobinopathy in northern Europe.²

To plan and provide adequate health care for patients with sickle cell disease it is essential to study the patterns of morbidity from sickle cell disease in the United Kingdom. The variability of the clinical course of sickle cell disease is well recognised and has been reviewed by Serjeant.⁷ Both genetic and environmental factors are known to determine the clinical severity. Detailed information about the genetic determinants such as the interaction with α thalassaemia, the presence of heterocellular high fetal haemoglobin, and the relevance of different haplotypes in different populations has become available.⁸⁻¹¹ In contrast the role of environmental factors, such as geographical and climatic effects, socioeconomic factors, quality of health care, the impact of education and information, etc, have not been quantified: longitudinal controlled data are not available.

In an attempt to establish the clinical characteristics of sickle cell disease in Britain we analysed all the acute admissions of patients with sickle cell disease to one district general hospital during one year and report our results.

Methods

A total of 221 patients with sickle cell disease were known to reside in the London borough of Brent on 1 September 1985, of whom 211 attended the department of haematology at this hospital for inpatient and outpatient care. Table I gives their ages and haemoglobin genotypes.

All of the patients were admitted under the care of the same team, to the

TABLE I—Patients with sickle cell disease who attended this hospital in September 1985

Genotype	Age (years)			Total No (n=211)
	Under 5 (n=32)	Between 5 and 16 (n=44)	Over 16 (n=135)	
SS	19	19	59	97
SC	11	18	54	83
SB*	1	2	6	9
SB+	1	5	12	18
Other*	0	0	4	4

*S-HPHF three, SJ one.

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TABLE II—Acute admissions for sickle cell disease, 1984-5. (Numbers of patients in parentheses)

Age (years)	Genotype			Total
	SS	SC	Other*	
<5	21 (10)	6 (3)	0	27 (13)
5-16	45 (12)	3 (3)	14 (3)	62 (18)
>16	60 (20)	7 (7)	5 (5)	72 (32)
Total	126 (42)	16 (13)	19 (8)	161 (63)

*Patients with SB^o and SB⁺.

TABLE III—Site of pain in 142 painful crises and duration of hospital stay according to site of pain and age (range in parentheses)

Age (years)	Limb	Trunk
Site of pain		
<5	12	7
5-16	23	32
>16	14	54
Duration of hospital stay (days)		
<5	5.4 (1-17)	8.6 (1-31)
5-16	4.9 (1-11)	8.3 (3-27)
>16	4.6 (1-9)	11.1 (4-35)
All	4.9 (1-11)	10.3 (1-35)

TABLE IV—Sequestration syndromes according to age

Age group	Sequestration syndrome			
	Splenic	Hepatic	Chest syndrome	Girdle syndrome
<5	1 (died)	1 (secondary to salmonella osteomyelitis)	—	—
5-16	—	—	3	1
>16	—	—	7	12 (one died)

same three wards (paediatric, adult male, or adult female), and treated according to a standard protocol of high fluid intake (oral or intravenous), antibiotics, and analgesia. As a part of the protocol various clinical investigations including microbiological studies were carried out. The microbiological investigations consisted of blood and urine cultures and swabs from the throat and other clinically indicated sites. Sera were collected during acute episodes and during convalescence for virological studies. The criteria for exchange transfusion were established in 1983 and followed throughout the study. Elective admissions for blood transfusion in individuals with strokes, renal failure, preparation for major elective surgery, and pregnancy were not included; there was a total of 99 admissions in this category, accounting for 293 hospital days.

Criteria for defining episodes of visceral sequestration were established and followed throughout the review (see below). The patients' records were reviewed by two of us (MB and SCD) independently and the results compared. The term vaso-occlusive crisis was used to denote all crises where the aetiology of presenting symptoms and signs as well as the subsequent course of the crisis was attributed to vaso-occlusion. Two types of vaso-occlusive crises were recognised: the painful crisis and other vaso-occlusive events such as strokes, priapism, and renal papillary necrosis. Painful crises were those where pain was the main presenting feature. Limb pain was defined as pain confined to one or more sites in the long bones, including the pain in the hip and shoulder joints, and trunk pain as pain in the vertebrae, pelvis, ribs, or abdomen, or in more than one site. If a patient had pain in the limbs as well as in the trunk this was considered as trunk pain.

A visceral sequestration syndrome was defined as a loss of function or acute enlargement of a visceral organ associated with severe systemic upset and a fall in haemoglobin, and often in the platelet count, from the steady state values. Four visceral syndromes were recognised: splenic sequestration as described by Topley *et al.*,¹² hepatic sequestration as described by Hatton *et al.*,¹³ the chest syndrome as reported by Davies *et al.* 1984,¹⁴ and "the girdle syndrome." The last syndrome was first recognised during a systematic study of abdominal pain in sickle cell disease (A Brownell *et al.*, in preparation) and is the most severe form of visceral sequestration in adults.

The syndrome consists of simultaneous or consecutive sickling and sequestration in the mesenteric circulation, the liver, and often the bases to midzones of the lungs. It is associated with pain in the lumbar spine, a rapid clinical deterioration, a silent distended abdomen, and reduced oxygen transfer. Patients with severe cases of the girdle syndrome require exchange transfusion.

Infective crises were defined as all episodes where an infectious agent was identified that could have contributed to the signs and symptoms. Three admissions were for reasons other than sickle cell disease: a drug overdose, a traffic accident, and a scalding accident.

Results

There were 161 acute admissions between September 1984 and September 1985 for 63 patients. Table II gives the patients' ages and genotypes. These patients spent a total of 1572 days in hospital—that is, 9.7 days per admission or 24.9 days for each patient admitted—a total of 7.4 inpatient days per patient with sickle cell disease in the district, which accords with our previously reported data.^{15 16}

One hundred and forty seven admissions (91%) were for vaso-occlusive events (142 for painful crises; three cerebrovascular accidents in patients aged 4, 4, and 18 years; two in pregnant women with renal papillary necrosis). Eleven admissions were for an infective or infection induced episode and three were for reasons unconnected with sickle cell disease. The incidence of trunk pain increased with age to become the predominant presentation site in patients over 16 (table III). Patients with limb pain stayed in hospital on average 4.9 days, whereas those with trunk pain remained for 10.3 days (table III).

There were 25 episodes of visceral sequestration, and these were seen only in the SS (14) and Sβ^o (one) patients. There were 23 episodes in

schoolchildren and adults presenting with trunk pain, one in a 4 year old with *Salmonella enteritidis* osteomyelitis, and one in an 18 month old baby newly arrived from Africa, who was not receiving prophylactic penicillin. Table IV gives details of the visceral sequestration episodes. Exchange transfusion was given in 12 episodes (eight patients: two chest syndromes, 10 girdle syndromes) and the patient with hepatic sequestration required supportive transfusions. Two patients died: the child with splenic sequestration, who was dead on arrival at the hospital, and a 19 year old boy with the girdle syndrome, who died despite intensive efforts at resuscitation. The youngest patient with the chest syndrome was 8 years old and with the girdle syndrome 13; the oldest patients with these syndromes in 1984-5 were 27 and 23 respectively.

Eleven infective episodes only were identified out of the 161 admissions (table V). All the patients under age 16 were taking prophylactic oral penicillin apart from the patient with splenic sequestration, who was mentioned above.

TABLE V—Infective episodes in sickle cell disease during one year

Age (years)	Genotype	Episode
0.5	SC	Viral illness; aplastic crisis
3.5	SS	<i>Haemophilus influenzae</i> meningitis
4	SS	<i>Salmonella enteritidis</i> osteomyelitis progressing to hepatic sequestration
13	SS	Rise in influenza A titres; painful crisis
13	SS	Parvovirus infection; aplastic crisis
14	SS	Rise in Q fever titres; painful crisis
14	SC	Mycoplasma pneumonia, parvovirus infection, aplastic crisis
16	SS	Parvovirus infection; aplastic crisis
20	SS	<i>Escherichia coli</i> ; urinary tract infection
25	SS	<i>Escherichia coli</i> ; urinary tract infection
29	SC	<i>Salmonella virchow</i> , splenic abscess

Discussion

Our observations confirm that over 90% of acute admissions in British patients with sickle cell disease are for painful vaso-occlusive crises. In nearly two thirds of all painful episodes the pain is in the trunk, and up to a quarter of such episodes culminate in severe visceral sequestration in the lungs, liver, or mesenteric circulation, or in more than one site. Young adults who experience visceral sequestration are severely ill, often requiring exchange transfusions and intensive support, and the mortality is considerable.

It is difficult to compare the clinical severity between different geographical locations. Recently, Brozović and Anionwu compared the clinical features of sickle cell disease in Brent and Jamaica and concluded that admission for painful crises was required by more patients with SS in Brent (92%) than in Jamaica (39%).¹⁶ Interestingly, patients with SS in Brent and Jamaica had a similar incidence of chest syndrome, priapism, splenomegaly, and aplastic crises. Leg ulcers, common in Jamaica, were rare in Britain. Patients with SC were less severely affected in Britain than in Jamaica. It is tempting to postulate that better health care in Britain during infancy and childhood results in the survival of most children born with SS, and those who escape the early mortality from pneumococcal infections and acute splenic sequestration grow up to suffer from frequent vaso-occlusive crises and die of the visceral sequestration syndromes. The natural history of sickle cell disease in a country with a mild temperate climate, liberal use of antibiotics, and accessible health care may be entirely different from that in the Caribbean or Africa and is probably closer to that seen in urban North America. Indeed, the severity of pain in the vaso-occlusive crisis in Britain and the USA requires parenteral opiates, whereas in Jamaica the pattern of pain is reputed to be less severe.^{17 18}

Our results confirm the need for planned health care for patients with sickle cell disease. Three important points arise:

(1) In Britain sickle cell disease is associated with an appreciable morbidity in older children, adolescents, and adults. Although the management remains primarily supportive and symptomatic, there is a need to develop a systematic approach to management, similar to the one recommended by the US Public Health Service.¹⁹

(2) The implications of the morbidity associated with sickle cell disease have predictable consequences for the use of acute beds. It can be calculated from our data that 50 patients with sickle cell disease will cause one hospital bed to be continuously occupied throughout the year with acute illnesses related to sickle cell disease. There are at least 20 health districts in Britain with 100 or more patients with sickle cell disease.²⁰ The needs of the patients in these

urban and often deprived districts must be taken into account when planning the acute services.

(3) Sickle cell disease may present as a medical emergency, and it is essential that guidelines for the management of the sequestration syndromes, cerebrovascular accidents, priapism, and infections are formulated, published, and implemented as soon as possible along the lines of the Department of Health guidelines for haemophilia and the US guidelines for sickle cell disease.²¹

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MATERIA NON MEDICA

First communes

The tiny sultanate of Brunei, enfolded by the east Malaysian states of Sarawak and Sabah, forms with them an irregular crescent over the vast, primitive, communist haunted (so it is said), bulk of Indonesia Kalimantan, the whole making up the island of Borneo. These beautiful lands of jungle, fast flowing rivers, golden domed mosques, Chinese temples, and street markets are home to a number of different peoples, which all keep their own culture, but live in apparent harmony. In six weeks of travel I never heard a voice raised in anger. As English is the second language, and widely spoken, visitors to Kuala Lumpur might consider venturing further afield than the obvious delights of west Malaysia and Thailand.

Borneo boasts two unique attractions—pitcher plants and longhouses. The plants, whether single—sometimes enormous specimens (one reportedly held a quart of water and a drowned rat)—or hanging epiphytically in tangled cascades, are indeed fascinating as are the less publicised ant plants. Both may be sought and unreservedly admired. Longhouses are different. They are homes, often of whole villages, and there is something distasteful about walking into somebody's home, even clutching the suggested bag of goodies. There is also the effect on the inhabitants, who have progressed, I was told, from generous hospitality through rightful suspicion (many

having been induced to part with valuable artefacts for a mere pittance) to something akin to begging. Tourists are now being urged further and further up the rivers to find unspoilt specimens.

I had signed on for a tour of the Niah Caves, themselves a wonder, both for their extent and the feats of the birds' nests harvesters 90 feet up on bamboo poles. In the event I was the only tourist, and Stanley, my guide, was much preoccupied with the problem of 19 German wildlifers who wanted not only to visit a longhouse but also to spend a night in one. Thus it was, after five hours of plank walking and cave crawling we set off longhouse visiting. I would gladly have put my feet up, but Stanley was my meal ticket. We visited four in all, that evening and the following morning. Two were dilapidated but with strong repairs to the building, one deserted (the inhabitants harvesting paddy, Stanley surmised) and one medium, 30 to 40 door sized. The communal area occupied about half the total, stretching further than the eye could see. It was beautifully clean, and the open doors let in pools of light in which sat groups of mothers, an old man mending a net, and a woman plaiting, while a youth drifted by, Walkman clamped in his ears. Side doors led to the private apartments of individual families, each apparently furnished with identical three piece suites.

The material advantages of such an arrangement in respect of shared overheads and mutual protection, are obvious. These structures were the first genuine communes, and the north Borneans, their head hunting past behind them, do seem to have achieved, to some extent, the submergence of self interest in the common good. Idea for export?—ANNE SAVAGE, London.