The causes and frequency of haemolysis in childhood vary according to age. In the neonatal period haemolysis is invariably; it may be physiological or pathological. Physiological haemolysis—destruction of red cells—is never more intense than in the haemolysis of fetal red cells just after birth, almost halving the initial circulating haemoglobin concentration by the time of resumption of erythropoiesis at about seven weeks after full term delivery. “Physiological” jaundice, resulting from this haemoglobin catabolism, is exaggerated to hyperbilirubinaemia in term babies by fetal and neonatal diseases, maternal disorders, or a combination of these. When there is a history of maternal smoking or administration of oxytocin the effects may be mediated through impairments in the baby’s red cell deformability, though the effects of oxytocin are controversial. Preterm infants are more at risk of haemolytic jaundice, not only because of hepatic functional immaturity but also because of their greater dependence at birth on mainly fetal red cells, whose viability is reduced for both functional and metabolic reasons.

Pathological haemolysis—haemolytic anaemia—is never more common than in the neonatal period. Underlying this may be intrinsic abnormalities of the red cell’s membrane such as hereditary spherocytosis, enzynopathies such as deficiency of glucose-6-phosphate dehydrogenase, or, rarely, even haemoglobinopathies such as homozygous sickle cell disease (though since this is a β globin chain abnormality it usually causes little haemolysis before the second month of life). Such haematological diagnoses of inherited red cell abnormalities may be made more confidently from the age of 3 months.

In white people, however, the most common cause of neonatal haemolytic anaemia is still fetomaternal isoimmunisation. Anti-Rhesus (Rh) D prophylaxis has reduced fetomaternal immunisation by almost 90%; in 1968 there were 750 stillbirths and infant deaths from Rh(D) haemolytic disease of the newborn, compared with 123 10 years later. Failure of anti-D prophylaxis may result from antenatal immunisation, inadvertent omission, or inadequate dosage. Antenatal anti-D prophylaxis may reduce maternal immunisation even further though the cost effectiveness of this remains controversial. With the success of anti-D prophylaxis, other antibodies are now becoming relatively more important. Any antibody which crosses the placenta may cause haemolytic disease of the newborn. As yet no other prophylaxis against maternal immunisation is available. After birth, phototherapy, transfusion of red cells, and exchange transfusion are still the mainstays of treatment of haemolytic disease of the newborn.

In older children acute autoimmune haemolytic anaemia causes haemoglobinuria and life threatening anaemia; acute autoimmune haemolytic anaemia is uncommon and so is unlikely to be seen more than a few times by any one haematologist or paediatrician, and their experience of treatment must be limited. The recent review by Sokol et al of 42 cases studied over the past 23 years at the Sheffield Regional Blood Transfusion Service, therefore, contains useful guidance in investigation, management, and prognosis of these rare but acutely worrying problems. Two thirds of the children were under 5 years old in whom the acute haemolysis usually occurred after trivial infections and was short lived. They recommend that steroids should be reserved for children suffering from persistent and severe haemolytic anaemia after two or three
weeks' supportive care. Splenectomy may very occasionally become necessary after lapse to chronic haemolyse in three to six months. Pneumococcal vaccination and oral penicillin minimise but do not abolish the infective hazards of hyposplenism. They found, as did others, that autoimmune haemolyse in childhood is only rarely associated with underlying disease such as systemic lupus erythematosus. This contrasts with experience in adults, in whom autoimmune haemolytic anaemia may be symptomatic of collagen diseases such as systemic lupus erythematosus or of lymphoma. The enigmas of the pathogenesis of autoantibody formation to red cells will be important to unravel. This diathesis may be partly genetically determined.

Recent progress has been made in the understanding and management of the haemolytic-uraemic syndrome, an uncommon but dramatic and life-threatening emergency. Its pathogenesis is now thought in some cases to hinge on imbalance between prostacyclin and thromboxane and on platelet-vascular interactions and endothelial damage. With greater understanding of the pathological basis of the haemolytic-uraemic syndrome, specific management may soon become available, at least in some cases. Evaluation of any proposed specific treatments would have to be adequately controlled or doubts will remain about their value, as in the recent report of management of epidemic and sporadic haemolytic-uraemic syndrome using vitamin E. Improvements in recent years in supportive care, particularly for renal failure, have improved the prognosis—especially of the sporadic form of the syndrome.

The common ground in management of these haemolytic anaemias acquired in childhood, whether caused by autoantibody formation or associated with metabolic and microvascular damage, is recognition of the importance of supportive, symptomatic care. If the child’s anaemia can be “tided over” for a week or two—and any associated disorder such as renal failure effectively managed—the only treatment needed may be red cell transfusions. More aggressive “haematological” treatments such as steroids, immunosuppression, or even splenectomy for immune-mediated haemolyse are indicated only with lapse over several weeks or months to chronic, severe autoimmune haemolytic anaemia.

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A chamber pot and Bible

Since 1981 seven young men have died of self inflicted injuries in the Glenochil complex in Scotland, which includes a detention centre and a young offenders institution, and at least three of them were deliberate suicides. After a fatal accident inquiry in 1984 into the death of William MacDonald, aged 17, who was found “hanging with a bedsheet round his neck in a half sitting position,” the Secretary of State for Scotland, Mr George Younger, set up a working party to review suicide precautions at the complex.

The working party was chaired by an Edinburgh forensic psychiatrist, Dr Derek Chiswick, and in something of a hurry it has produced a radical document that has implications far beyond Glenochil. More information on the working party’s 63 recommendations is given on p 353, but here we want to highlight the recommendation to abolish what is called “strict suicide observation.” This observation is applied to inmates thought to be at risk of suicide. They are admitted to special cells in which all the washbasins, fitted furniture, and protruding fitments have been removed; an electric light, operable from the outside, burns all the time, and the room contains only a desk and chair made from toughened cardboard, a plastic chamber pot, a paperback book (or a comic), and a copy of the Bible. During the day the inmate has one blanket made of coarse, untearable canvas, and at night he is given another such blanket and a mattress; during the winter, staff and inmates agreed, the cells can be extremely cold. The inmate usually has to wear a canvas gown and is not allowed underclothing or any other clothes. He sits in his room all day, is observed every 15 minutes, and is not permitted any association with other inmates. In 1984, 164 inmates were placed on strict suicide observation for between two and 365 days, and four were there for more than nine months.

The working party describes this regimen as “inhumane and unacceptable,” condemning the denial of human contact as “contrary to modern notions of psychiatric care.” It has suggested instead a system more like that used in psychiatric hospitals that depends on human contact and team work from nurses, psychiatrists, and psychologists. The Secretary of State has accepted “in broad outline” the new system but has refused to abolish strict suicide observation on the grounds that such a move would have to be approached cautiously and examined for the whole prison system (adult as well as young offender). This seems to us not an argument for rejecting the recommendation for Glenochil but rather for applying it to the whole of the British prison system.