The weights of the 25 preterm infants ranged from 800 to 2550 g (mean 1411 g) and gestation from 27 to 36 weeks (mean 30.7 weeks). Eight infants had been ventilated in the neonatal period for a maximum of 13 days; two infants had suffered intraventricular haemorrhage; and one had received an exchange transfusion. None had suffered from necrotising enterocolitis or any other abdominal pathology and none had developed chronic lung disease. In the whole group there were six twins—five boys and one girl, but no pairs of twins, although two of a set of male triplets were included.

Discussion

In this study 39% of all herniotomies under 6 months of age were carried out on preterm infants. We have shown a 10 fold increase in risk in infants born before 36 weeks' gestation and a 20 fold increase in very low birthweight infants. This is in keeping with the findings of other workers who have reported an incidence of up to 30% in very preterm infants.¹ It seems that larger preterm infants are also at risk. There has been disagreement as to whether the increased risk applies equally to both sexes—this study suggests that this is the case.

Persistent patency of the processus vaginalis or canal of Nuck is considered important in the development of inguinal hernia but as it is found in over 90% of all newborns² there must be additional factors operating in the preterm infant. Keith³ has suggested laxity of the inguinal ring as a factor. We speculated that increased intra-abdominal pressure due to problems such as necrotising enterocolitis, feed intolerance, or chronic lung disease could be

important. Our findings did not support this hypothesis.

This study confirms that inguinal hernia is a common complication in preterm boys. The risks of operation in a small infant who may have residual lung disease are obvious. For this reason treatment is often delayed, especially in areas where there is no ready access to specialist paediatric surgeons and anaesthetists. Complications, however, such as incarceration and strangulation are especially common in young infants and delay may have tragic consequences.⁴ It is important that doctors involved in the early care and follow up of preterm infants are vigilant. We would advise early operation, preferably by a specialist paediatric surgeon. If the diagnosis is made on the neonatal unit, operation before hospital discharge is recommended.

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Recurrent infections with IgG₂ deficiency

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SUMMARY An 11 year old girl with retarded growth, recurrent infections, bronchiectasis, and normal serum immunoglobulin concentrations had a combined deficit of the IgG₂ subclass and IgG and IgM specific antibodies. Immunoglobulin replacement was followed by clinical improvement. The importance of determining both IgG subclasses and antibody activity in patients with recurrent infections and normal serum immunoglobulin values is emphasised.

Normal concentrations of serum immunoglobulins in children being evaluated for recurrent infections are generally accepted as evidence of intact humoral immunity. Immunoglobulin concentrations, however, may be normal in the presence of subclass deficiency, as recently described for IgG₂ deficiency¹ or antibody deficiency, or both.² We describe an 11 year old girl who presented with recurrent respiratory tract infections, normal serum immunoglobulin concentrations, and a combined deficiency of IgG₂ and specific IgG and IgM antibodies.

Case Report

An 11 year old girl was admitted to hospital in 1982 for immunologic evaluation (Table) because of repeated episodes of conjunctivitis, otitis media, and pneumonia beginning in the first year of life. Sputum cultures had shown *Pseudomonas* and *Candida* on several occasions during previous hospital admissions for pneumonia.

On this occasion she was below the third centile for height and weight and had conjunctivitis and clubbing of the fingers. The spleen was palpable 1 cm below the left costal margin. Chest radiographs suggested bilateral bronchiectasis which was confirmed by bronchography. Full blood count and a sweat chloride test were normal (Table). The patient had low serum IgG₂ subclass values together with a deficiency of antitetanus, anti-*Escherichia coli* specific IgG, and antirabbit red cell IgM antibodies. In addition, isohaemagglutinins were present in very low titres. Tetanus toxoid booster immunisation resulted in only a very low antibody response. These findings, together with the data indicating normal cell mediated immunity, were considered evidence of abnormal humoral immunity warranting replacement treatment with standard

human serum globulins. Subcutaneous treatment with human serum globulins (100 mg/kg per week) was started and was followed by a decrease in the frequency of infections. Activity of the alternative pathway of complement, which was impaired at the time of initial evaluation, returned to normal after the first few months of replacement treatment, as previously reported in patients with agammaglobulinaemia.³ The serum IgG value also increased to 120 IU/ml and the percentage of IgG₂ became normal (45% of normal pooled sera). In addition, the titres of antibodies to *E coli* and tetanus toxoid increased from 0.8 to 100 arbitrary U/ml and from 0.3 to 4 µg/ml, respectively.

Discussion

Selective deficiency of IgG subclasses has been reported as a cause of recurrent pyogenic infections with *Haemophilus influenzae*, *Streptococcus pyogenes*, and *Staphylococcus aureus*.² Recently Bass *et al*¹ reported a case of recurrent meningococcaemia associated with low IgG₂ concentrations, but normal values of the total serum immunoglobulin classes. The child described here presented with recurrent respiratory infections from the first year of life, eventually leading to bronchiectasis. Humoral immunity, as judged by serum immunoglobulin values and secretory IgA, was seemingly normal. Determination of IgG subclasses, however, showed a low IgG₂ value. Furthermore, a low specific IgM response, as judged by the low isohaemagglutinin and antirabbit red cell titres, was observed together with a low IgG antitetanus response. IgG antitetanus antibodies belong mainly to the IgG₁ subclass.⁴ So called antibody deficiency with normal immunoglobulins has been reported in several patients,⁵ but simultaneous assessment of IgG subclasses has seldom been carried out. Antibodies to several antigens have been found to be low in many patients with selective subclass deficiency.⁶ Together with the present report, these observations suggest that 'antibody deficiency with normal immunoglobulins' and 'IgG subclass deficiency' may represent overlapping syndromes. In fact in our patient, the IgG₂ deficiency did not account for the antibody deficiency, which included IgM as well as antibodies normally belonging to the IgG₁ subclass. These findings emphasise the importance of measuring both IgG subclasses and specific antibody activity when evaluating humoral immunity.

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Table Results of immunologic studies at time of diagnosis

	Patient's values	Normal values [†]
White cell count (×10 ⁹ /l)	7	5.5–10.8
Circulating neutrophils (×10 ⁹ /l)	3.5	3–7
Circulating lymphocytes (×10 ⁹ /l)	1.8	1.5–4.5
Serum immunoglobulins (IU/ml):		
IgG	96	88–238
IgA	60	42–270
IgM	274	72–323
Serum IgG subclasses (% of normal pooled sera)		
IgG ₁	150	88–126
IgG ₂	20	36–126
IgG ₃	170	17–128
IgG ₄	20	5–375
Salivary IgA	Present	Present
Antibody activity		
IgG: antitetanus (µg/ml): Baseline	Undetectable	Undetectable
After booster	0.3	2–8
Anti- <i>Escherichia coli</i> (AU/ml)	0.8	≥30
IgM: isohaemagglutinins*: Anti-A	1/2	≥1/64
Anti-B	1/2	≥1/64
Antirabbit red cells	Undetectable	≥128
Complement activity (HU/ml):		
Classic pathway	1000	900–1400
Alternative pathway	32	63–132
Lymphocytes forming E rosettes (%)	65	50–75
In vitro lymphocyte response to (counts/min)		
Phytohaemagglutinin	58 000	≥50 000
Concanavalin A	12 200	≥10 000
Pokeweed mitogen	5420	≥5000

* Blood group O.

[†] Normal values for age according to our laboratory.

AU=arbitrary units.

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Parents' understanding of coeliac disease and diet

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SUMMARY Parents of 50 children with coeliac disease were questioned about their knowledge of the disorder and the child's dietary compliance. Twenty of 50 parents admitted that compliance was less than strict. This correlated with poor knowledge of the disease which in turn was related to the family's social status. Membership of the Coeliac Society (23 of 50) correlated with both parental understanding and dietary compliance. Greater emphasis on dietary compliance and education of parents and patients is required.

Childhood coeliac disease requires a strictly kept gluten free diet for initial catch up growth and long term health and prevention of nutritional deficiencies¹ and possibly malignancy in later life.²⁻⁴ Few dietary studies have been conducted into childhood coeliac disease, and none into parent's understanding of the disorder or the effort directed towards dietary compliance.

Patients and methods

Fifty children (29 girls) with coeliac disease were studied at 1.5 to 19 (median 9.9) years of age. The diagnosis had been made on the basis of severe villus atrophy on jejunal histology and good clinical response to a gluten free diet.⁴ Where these two criteria applied no further biopsies were done. Patients and parents had been counselled by medical and dietetic staff both at diagnosis and at regular long term review.

A questionnaire was given to the parent who attended with the patient for clinical review. Questions were designed to test knowledge of the disease and the gluten free diet, probe attitudes and motivation, and determine the degree of dietary compliance. The family's social index⁵ was determined from information about parental education, work, housing, and family size. A clinical examination including measurement of height and weight was carried out, blood samples were taken to assess nutritional indices, and clinical information was abstracted from the patient's past medical records.

Results

Twenty nine families belonged to the registrar general's social class IV or V, 21 to class II or III. Only five parents had attained the GCE ordinary level examination standard or higher.

Dietary compliance. Parents were asked whether the diet was being kept and were given three options—strictly maintained (30 patients); broken regularly or sometimes (18); diet kept poorly or ignored (2). Dietary compliance was unrelated to the child's sex or current age, age at diagnosis, time since diagnosis, presence of a family history of coeliac disease (9 children), or presence of only one parent at home (11 families). Before the beginning of this study, eight patients had required a 'compliance' biopsy because of serious doubts about dietary compliance, and six of these admitted during the study that dietary compliance was still less than strict ($P < 0.07$).