In most, but not all, cases of neonatal myotonic dystrophy, the mother of an affected child will be found at some stage of the child's life to have myotonic dystrophy herself. Harper and Dyken (1972) report on 40 children with myotonic dystrophy during the first 5 years of life. In 4 of these children the onset was during the neonatal period. If respiratory failure in a newborn infant is due to myotonic dystrophy it is extremely likely, though not inevitable, that the mother of the child will also have the features of the condition.

Death from myotonic dystrophy in the neonatal period is uncommon. It would be helpful to know of any case which had survived involvement as severe as that recorded here.

**Summary**

Myotonic dystrophy should be included in the differential diagnosis of neonatal respiratory failure accompanied by hypotonia. The effect of this disorder in an infant who died from it 49 hours after birth is described, and the importance of examining the mother of a possible case is emphasized.

**REFERENCES**


**Effect of thermal environment and caloric intake on head growth of low birthweight infants during late neonatal period**

It has been shown that exposure of low birthweight infants to environmental temperatures slightly below the thermoneutral zone is associated with decreased rates of weight gain and linear growth (Glass, Silverman and Sinclair, 1968, 1969). It has been suggested that a combination of low environmental temperature and suboptimal caloric intake may be responsible for decreased rates of head growth (Davies and Davis, 1970; Glass, Silverman, and Sinclair, 1971), and by inference, brain growth (Winick and Rosso, 1969).

In the present study, matched low birthweight infants were reared under one of 4 combinations of thermal environment and caloric intake after the first week of life. The retarding effect of the subthermoneutral temperatures on head growth was confirmed.

**Subjects and methods**

Forty-two asymptomatic neonates (birthweight 930–1800 g), matched for birthweight and gestational age, were included in the study (Table I). During the first

**TABLE I**

*Characteristics of infants in study*

<table>
<thead>
<tr>
<th>Group</th>
<th>No. of infants</th>
<th>Birthweight (kg) (mean and range)</th>
<th>Gestational age (w) (mean and range)</th>
<th>Head circumference (cm) (mean and range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>11</td>
<td>1.60 (1.16–1.80)</td>
<td>35 (32–36)</td>
<td>29.2 (27.5–31.6)</td>
</tr>
<tr>
<td>II</td>
<td>10</td>
<td>1.62 (1.32–1.77)</td>
<td>35 (31–38)</td>
<td>30.0 (27.5–31.5)</td>
</tr>
<tr>
<td>III</td>
<td>10</td>
<td>1.60 (0.93–1.80)</td>
<td>34 (32–37)</td>
<td>29.4 (26.0–31.5)</td>
</tr>
<tr>
<td>IV</td>
<td>11</td>
<td>1.62 (1.12–1.80)</td>
<td>34 (30–36)</td>
<td>28.9 (27.1–31.5)</td>
</tr>
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</table>


**KENNETH SIMPSON**

Leicester General Hospital, Leicester.

*Correspondence to Dr. K. Simpson, The Leicester Royal Infirmary, Leicester LE1 5WW.*
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Exposure to slightly cool environmental temperatures may cause diversion of calories intended for brain growth into 'fuel' for heat production.

Summary

In order to assess the effects of ambient thermal conditions on postnatal head growth in low birthweight infants, 42 asymptomatic neonates were reared under 4 combinations of caloric intake and thermal environment after the first week of life. Exposure to a subthermoneutral temperature (abdominal skin temperature of 35°C), together with a relatively low caloric intake (120 cal/kg per day), was associated with significant retardation of head growth over a 2-week study period.

References


Simultaneous occurrence of diabetes, liver cirrhosis, and 47, XX, 21+ /46, XX chromosomal pattern

There is an unduly high incidence of diabetes among patients with Down's syndrome (Burch and Milunsky, 1969; Serrano-Rios et al., 1973), and also of autoimmune processes (Burgio et al., 1965; Harris and Koutsoulieris, 1967). There also appears to be some connexion between liver cirrhosis and diabetes (Creutzfeldt, Wille, and Kaup, 1962), but the simultaneous occurrence of these two conditions in a patient with a 21-chromosome abnormality has not been previously reported.

Case report

The female patient had been admitted first at the age of 6 years with stunted growth, upper respiratory infections, urticaria and other allergic symptoms, and an enlarged liver. Liver function tests had been abnormal; serum IgG level was raised.

Her diabetes became manifest at the age of 8 years, since when she has been on insulin treatment. The diabetes was labile, and hyper- and hypoglycaemic episodes have occurred frequently. At that time the thymol turbidity test was 12–18 units; SGOT about 100 IU; serum bilirubin level normal. She had never been jaundiced. Tests for Australia antigen negative. Direct antiglobulin test negative. Tests for antibody formation proved normal.

Reinvestigated at 18 years, her development was infantile, height only 134 cm, weight 36 kg. The liver reached the umbilicus. The IgG level was raised (Table).

<table>
<thead>
<tr>
<th>TABLE Serum protein fractions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before onset of diabetes</td>
</tr>
<tr>
<td>Total protein (g/100 ml)</td>
</tr>
<tr>
<td>Albumin (%)</td>
</tr>
<tr>
<td>Serum electrophoresis</td>
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<tr>
<td>α1 (%)</td>
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<td>α2 (%)</td>
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<td>β (%)</td>
</tr>
<tr>
<td>γ (%)</td>
</tr>
<tr>
<td>Immune electrophoresis</td>
</tr>
<tr>
<td>Prealbumin</td>
</tr>
<tr>
<td>Albumin</td>
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<tr>
<td>βγ complement</td>
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<td>IgA</td>
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<td>IgM</td>
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<td>IgG</td>
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</tbody>
</table>

Slightly decreased —, decreased — —, considerably decreased — — —. Slightly increased +, increased + +, much increased + + +.

Lymphocyte culture showed a 47,XX,G+ pattern in 20 cells of the 208 cells examined. The same 47,XX,G+ karyotype was seen in 2 out of 30 bone marrow cells. Giemsa staining showed the extra chromosome to belong to the 21 pair. On the other hand, in a fibroblast culture all the 97 cells examined yielded 47,XX pattern. The karyotype of the parents was normal. In view of these findings, bone marrow, liver, and spleen biopsies were carried out.


Departments of Pediatrics, Jewish Hospital and Medical Center of Brooklyn, and Harlem Hospital Center, New York City, U.S.A.

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L Glass, R V Lala, V Jaiswal and S K Nigam

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