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.

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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

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

week of life, all of the infants were reared nude except for a napkin in servocontrolled Isolette incubators with abdominal skin temperatures maintained at 36.5°C (a condition associated with thermoneutrality). Feeds of Similac with iron (0.8 cal/ml) were started within the first 8 hours of life. These were supplemented with intravenous infusions of 10% glucose solution until a daily caloric intake of at least 80 cal/kg per day was tolerated orally.

When an oral caloric intake of at least 120 cal/kg per day had been attained, usually by the end of the first week, the infants were randomly assigned to one of 4 study groups: I, abdominal skin temperature 36.5°C, caloric intake 144 cal/kg per day; II, abdominal skin temperature 36.5°C, caloric intake 120 cal/kg per day; III, abdominal skin temperature 35.0°C, caloric intake 144 cal/kg per day; IV, abdominal skin temperature 35.0°C, caloric intake 120 cal/kg per day. Abdominal skin temperatures of 36.5°C are associated with incubator air temperatures of approximately 34.5°C in these infants, while skin temperatures of 35.0°C are associated with incubator air temperatures of approximately 32.0°C (Glass *et al.*, 1968). Infants who were small-for-gestational age (birthweight between the 3rd and 10th centiles) were divided equally among the 4 groups. While there was slight intergroup variation in gestational age, initial head circumference was similar in all groups.

Each infant was weighed before entering the study and daily thereafter by the same group of nurses before the first morning feed. Weights were recorded to the nearest 10 g. Volumes of feeds were adjusted after weighing infants to provide each with the number of calories specified by the protocol.

Head circumference was measured (to the nearest 0.1 cm), using a K & E Whyteface steel measuring tape, on admission to the study, twice weekly, and on completion of the trial. Rates of weight gain were expressed as the daily rate of increase (percentage of current weight per day) and head circumference as the average daily increment in cm/day. Intergroup comparisons were made by the 2 standards ranks test of Wilcoxon and White (Mainland, 1963).

Results

The mean and range of daily rates of increase in weight and head circumference are shown in Table II. The differences in weight gain between both 'warm' groups (I and II) and group IV were of borderline significance ($0.1 > P > 0.05$) for each comparison. While the infants in group I gained weight more rapidly than group II infants, no significant differences were observed. There was no significant difference between the rate of weight gain of group III infants and those of either groups I and II (the warm groups) or group IV.

The differences in daily increase in head circumference between both 'warm' groups and group IV were significant (for I *v.* IV, $P < 0.05$; for II *v.* IV, $P < 0.01$). The daily increment in the

TABLE II
Daily rates of increase in weight and head circumference

Group	Rate of weight gain (%/d) (mean and range)	Rate of increase in head circumference (cm/d) (mean and range)
I	1.68 (1.02–2.66)	0.15 (0.09–0.20)
II	1.51 (1.06–2.16)	0.15 (0.11–0.18)
III	1.46 (0.78–2.25)	0.14 (0.07–0.18)
IV	1.25 (0.90–1.61)	0.13 (0.06–0.20)

circumference in group III was intermediate between group IV and the 2 'warm' groups, and not significantly different from either.

Discussion

It has been known for over a decade that rearing low birthweight infants in a thermoneutral environment during the early neonatal period is associated with increased survival rates (Buetow and Klein, 1964; Day *et al.*, 1964), but optimal thermal environment after the first days of life has never been clearly established.

In the present study we have demonstrated several points. Environmental air temperatures of about 32°C, which are associated with an increase in resting metabolic rate of about 20% in nude infants of low birthweight after the first week of life (Glass *et al.*, 1969), have a retarding effect on head growth. This is partially compensated for by an increase in caloric intake. In addition, there does not appear to be any advantage in increasing the daily caloric intake above 120 cal/kg per day, provided the infant is not exposed to slightly cool environmental temperatures.

We must assume that the relatively slow rate of head growth in the group IV infants was associated with decreased brain growth during this critical period of life (Winick and Rosso, 1969).

While the mean difference in increase in head circumference between group IV and groups I and II, the warm groups, was only about 0.3 cm over the 2-week study period, it must be remembered that the volume of cranial contents is directly proportional to the cube of the head circumference.

It has not yet been determined whether these differences in head growth over this short (but very critical) period of time ultimately affect neurological development. However, it seems wise at this time to rear low birthweight infants in a thermoneutral environment after the first week of life, since ex-

posure 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 birth-weight 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.

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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 12–19 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
Serum protein fractions

	Before onset of diabetes	After onset of diabetes	After prednisolone treatment
Total protein (g/100 ml)	10.8	9.7	6.4
Albumin (%)	35	24	35
Serum electrophoresis			
α_1 (%)	3	5	6
α_2 (%)	7	12	21
β (%)	10	13	18
γ (%)	45	46	20
Immune electrophoresis			
Prealbumin		— —	Normal
Albumin		— — —	—
β_2 C complement		+	+
IgA		+	+
IgM		++	++
IgG		+++	Normal

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.