

that has not previously been shown. It is surprising that the serum complement level has remained normal.

The thickening of capillary walls is seen on electronmicroscopy to be due to subendothelial aggregations and increase in cytoplasm, but there is not the marked 'layering' present, which is seen in membranoproliferative glomerulonephritis (Cameron *et al.*, 1970). Though one of the two isolated electron dense aggregations, illustrated in Fig. 2, appears to project from the epithelial surface it is not typical of a 'hump' as seen in poststreptococcal glomerulonephritis (Herdson, Jennings, and Earle, 1966). It is separated from the podocytic cytoplasm by a fine layer of lamina densa, unlike a hump which rests on the epithelial side of the basement membrane totally surrounded by fused foot processes.

The finding of staphylococcal antibodies in the blood and nodular deposits of staphylococcal antigen together with complement, fibrinogen, and IgG in the glomeruli, supports the concept of an immunological cause of the glomerulonephritis. The electron dense aggregations in the basement membrane most resemble the nonspecific aggregations found in acute poststreptococcal glomerulonephritis and are considered to be antigen-antibody complexes (Herdson *et al.*, 1966). The glomerulonephritis associated with *Staph. albus* infection of an atrioventricular shunt appears to be a further example of immune complex disease in man similar to that found in experimental glomerulonephritis in animals (Dixon, Feldman, and Vazquez, 1961).

Summary

A 3½-year-old mentally retarded boy who had had a Spitz Holter valve inserted at 10 months for hydrocephalus developed the nephrotic syndrome with haematuria, acidosis, and uraemia. *Staphylococcus albus* was cultured from CSF obtained from the valve. Antibody to *Staph. albus* was present in the blood at a titre of 1/5120, but serum β_1 C globulin was normal. The infected valve was removed, and he was treated with antibiotics for 3 months. The nephrotic syndrome has remitted and renal function has returned to normal. Renal biopsy showed proliferative glomerulonephritis with variable thickening of the capillary walls, and electronmicroscopy showed marked thickening of the basement membrane due to subendothelial deposits.

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Muscular performance in cystic fibrosis patients and its relation to vitamin E

It is generally felt that children suffering from cystic fibrosis (CF) should be encouraged to lead as normal lives as possible and this in particular includes full physical activity. It was our clinical impression that many CF children performed extremely well at various sports and we therefore wished to measure their muscle performance in carefully controlled tests as compared with normal subjects.

In addition, there is current interest as to whether vitamin E may play a role in athletic performance (*British Medical Journal*, 1971), and since cystic fibrosis sufferers have consistently low vitamin E blood levels (Bennett and Medwadowski, 1967; Muller and Harries, 1969; Harries and Muller,

TABLE I
CAHPER* Fitness—Performance Tests used in trial

Test	Purpose	Method	Result
Flexed arm hang	To test muscular endurance of arm and shoulder muscles	Subject hangs with reverse grip from a bar, keeping eyes above, or at bar level	Longer time hanging, of two trials, is recorded
Standing broad jump	To measure explosive strength (power) of legs	Subject jumps forward from standing position	Distance achieved in the better of two trials is recorded
One-minute speed sit-up	To test muscular strength and endurance of abdominal muscles	From supine position subject sits up, touching both elbows to flexed knees, and then returns to starting position	Number of sit-ups done in 1 minute is recorded
Shuttle run	To test agility	Starting from prone position child runs 30 ft to collect 2 wooden blocks (one at a time) and return them to behind starting line	Time taken in better of 2 trials is recorded

*Canadian Association for Health, Physical Education and Recreation.

1969), we wished to determine whether their muscle performance was reduced.

Subjects and methods

Muscle testing was performed on 20 children whose diagnosis of cystic fibrosis had been confirmed by finding raised sweat sodium values (sweat sodium >70 mEq/l. in at least 100 mg sweat). Our group consisted of 12 boys, aged 7 to 16 years, and 8 girls, aged 8 to 14 years. These children were chosen because they lived within easy reach (30 miles) of the testing centre and were within the age range for which we had control standards for muscle testing. No child from our clinic list was excluded on physical grounds. All the children except Case 1, were prepubertal. The children were seen in groups of four in the outpatient department and all the testing was carried out between 10 a.m. and midday.

The muscle tests used were taken from the *CAHPER Fitness-Performance Test Manual* (Hayden and Yuhasz, 1966) and each one used measured a different aspect of fitness. They were performed in the recommended manner, explained in Table I.

We also tested hand-grip strength. The children squeezed a specially designed, compressible bulb, blown up to 40 mmHg and in continuity with a graduated mercury column. This has previously been used by Carthum, Clawson, and Decker (1969). The highest pressure achieved in 3 tries for each hand was recorded. Values for each age and sex had been previously obtained from normal schoolchildren in the Vancouver area (R. H. Hill, personal communication, 1971).

The tests were done in a standard order and the children encouraged to give their best performance.

All children had tests of respiratory function and chest *x*-ray (PA and lateral) before the fitness-performance assessments. (The respiratory function tests were performed as a baseline for a further study to assess the effect of vitamin E in this group and will be reported later.)

The chest *x*-ray was scored for each child according to the system developed at The Hospital for Sick Children, Great Ormond Street, London (A. P. Norman and A. R. Chrispin, personal communication, 1971). This assesses chest configuration, pulmonary shadows due to bronchial wall thickening, small areas of lobular consolidation, alveolar wall thickening and breakdown, segmental or lobular collapse and/or consolidation, and degree of pulmonary artery unfolding. Each category is scored not present (0), present but not marked (1), and marked (2), giving a possible score range 0–40.

Shwachman scoring (Shwachman and Kulczycki, 1958) was also done for each child.

Blood was taken at least four hours p.c. for vitamin E estimation, performed according to the Emmerie-Engel colorimetric method of Quaife and Harris (1944).

Results (see Table II)

The patients in the Table II are in order of merit on Shwachman score. It can be seen that they ranged from excellent condition to those moderately affected by disease.

The chest *x*-ray scores correlate well with the Shwachman score ($r = -0.81$). Those patients with mild to moderate disease on clinical scoring in general have moderate to extensive changes of disease on chest *x*-ray.

The results of the CAHPER muscle performance tests are expressed as centiles for that age and sex, and the hand-grip strength results are expressed in quartiles.

Plasma vitamin E results are in the right-hand column. All the values are extremely low as compared to those obtained for healthy subjects (Desai, 1968), with the exception of Case 19 who has little bowel disturbance and a vitamin E level in the lower range of normal.

The vitamin E levels in cystic children ranged

TABLE II

Results

Shwachman grading	Case no.	Age	Sex	Shwachman score	Chest x-ray score	Handgrip (quartile)	Sit-up (centile)	Broad jump (centile)	Shuttle run (centile)	Arm hang (centile)	Plasma vitamin E (mg/100 ml)
Excellent	1	13	F	95	3	> 25	50	80	75	> 50	0.15
	2	11	M	90	4	50	65	80	40	> 85	0.05
	3	7	M	90	8	> 25	> 30	90	15	> 80	0.22
	4	8	M	84	1	> 50	40	15	> 10	50	0.01
	5	12	M	82	4	> 25	0-5	> 15	> 35	> 15	0.09
Good	6	15	M	79	7	< 25	35	> 15	15	> 25	0.43
	7	10	F	78	3	50	75	65	> 30	> 90	0.13
	8	15	M	77	8	25	40	20	25	85	0.41
	9	8	F	75	7	> 25	55	> 75	> 55	15	Parents refused blood test
	10	7	M	74	7	50	Not done	15	> 15	15	0.16
Mild	11	9	M	69	10	> 25	5	60	> 10	> 70	0.08
	12	11	F	67	7	> 25	Not done	45	> 20	> 5	0.14
	13	12	M	64	8	< 25	> 85	60	> 75	> 65	0.02
	14	10	F	64	9	< 25	> 90	70	> 40	> 30	0.09
	15	8	F	61	13	> 25	85	> 65	> 5	> 85	0.45
Moderate	16	16	M	59	11	> 25	45	55	30	> 35	0.09
	17	13	F	59	12	< 25	> 95	> 35	> 15	80	0.44
	18	14	F	58	6	< 25	65	> 45	> 35	> 70	0.34
	19	7	M	54	12	< 25	35	> 40	0-5	> 70	0.74
	20	12	M	44	16	< 25	> 85	90	95	> 90	0.07

from 0.01 to 0.74 mg/100 ml (mean 0.22 ± 0.13 mg/100 ml) as compared with mean values of 0.90 ± 0.22 mg/100 ml for healthy subjects under 25 years (Desai, 1968).

Discussion

The children were evenly distributed between Shwachman 'excellent to good' and 'mild to moderate' categories of disease.

In the muscle performance tests, all children did within normal standards except for two children in one family (Cases 12 and 10) who did not perform sit-ups. We felt this was not due to physical weakness but rather to a familial attitude.

The test performed least well by the group as a whole was the shuttle run, for which we have no satisfactory explanation. In the CAHPER tests there was no correlation between performance and clinical status in that 17 of the 38 tests scored at the 50th centile or above were by patients in the excellent to good clinical category and the remaining 21 were by those in the mild-to-moderate group. Of the 42 tests scored below the 50th centile, 23 were from the excellent-to-good patients and 19 from the mild to moderate category. In the test of hand-grip strength, however, 7 children scored below the normal lower quartile, 6 of whom were in the mild to moderate category of disease.

These results show that children with cystic fibrosis are able to perform well in tests of muscle power, endurance, and agility. They support our clinical impression that these children often reach

above average athletic standards despite respiratory problems.

The results of vitamin E estimation in this group of cystic fibrosis children compare excellently with previously published results (Harries and Muller, 1969). There was no correlation between actual vitamin E blood level and skeletal muscle function and it is apparent from our study that low blood vitamin E levels did not prevent our subjects achieving normal standards in these tests. From this preliminary trial, it is impossible to tell whether these children's performances would improve on raising their blood vitamin E level towards normal. For this reason, a controlled trial of vitamin E supplementation is now in progress on this group and will be reported later.

Summary

Tests of muscle power and endurance were performed on 20 children suffering from cystic fibrosis. The results compared well with standards of normal Canadian schoolchildren and many cystics performed well above average, despite moderately severe respiratory disease, as judged by clinical and chest x-ray scoring. 18 of the 19 children tested had an extremely low blood vitamin E level. There was no correlation between the vitamin E level and skeletal muscle function.

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