MUSCLE DISORDERS IN CHILDHOOD

Volume XIV in the series
MAJOR PROBLEMS IN CLINICAL PEDIATRICS

DUBOWITZ

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Dedicated to

DAVID SMITH (1943–1959)

He had the intellect to understand
and the courage to accept

Foreword

Muscle Disorders in Childhood covers a broad spectrum of diseases which plague the pediatrician because of the complexity of their pathogenesis and the difficulty of their differentiation. Victor Dubowitz, Professor of Pediatrics at the Institute of Child Health and the Royal Postgraduate Medical School of the University of London, and Honorary Consultant Pediatrician to the Hammersmith Hospital, has attacked this problem and clarified it in this latest addition to our series, *Major Problems in Clinical Pediatrics*.

Victor Dubowitz, a graduate in medicine of the University of Cape Town and PhD of the University of Sheffield, is known to the American medical public as the father of the Dubowitz scoring system which calculates the newborn's gestational age at birth. He is indeed the father, but the scoring system has a mother too. She is, quite appropriately, his wife, Lilly Dubowitz. But Dr Dubowitz's fame rests even more securely upon his contributions to knowledge concerning the muscular dystrophies and atrophies, with special reference to the histological and histochemical alterations in these disorders. He has devoted the greater part of his professional life to the clinical, pathological and molecular study of normal muscle and its diseases.

He has written four books, *Developing and Diseased Muscle: A Histochemical Study*; *The Floppy Infant*; *Muscle Biopsy: A Modern Approach*; and *Gestational Age of the Newborn: A Clinical Manual*. He has lectured widely at scientific meetings all around the world, from Aberdeen to Zürich, from Australia to the United States, and has made many original contributions to medical journals. *Muscle Disorders in Childhood* presents a synthesis of the advances in knowledge in the clinical and pathological aspects of muscle disease over the past 20 years. We venture to guess that he is the most appropriate man in the world to undertake this formidable task. In our opinion, he has succeeded admirably.

ALEXANDER J. SCHAFFER, MD

Preface

The past few years have seen an avalanche of interest in muscle disorders, coming from various directions – clinicians, pathologists, biochemists, geneticists, cell biologists, physiologists and a host of other scientists. Although we still remain ignorant of the cause of most of the major genetic disorders of the neuromuscular system, important strides have been made in various areas of study. It thus seemed timely to try and bring together the clinical and associated advances.

My interest in muscle disorders began in 1957 during a residency at Queen Mary's Hospital for Children in Carshalton, Surrey. Part of my duties entailed an occasional call to the Muscular Dystrophy Ward to treat a child who had pneumonia or other severe illness. Having never heard of muscular dystrophy, let alone seen a case, my curiosity was fired and from this clinical beginning grew an interest in investigating this mysterious condition. This led me to the laboratory of Professor Everson Pearse at the Royal Postgraduate Medical School at Hammersmith Hospital, where I spent many an interesting evening (or night) applying the then relatively new techniques of histochemistry to the biopsies I had taken from the patients.

My fruitful association with Queen Mary's Hospital for Children and the Royal Postgraduate Medical School reinforced my view that the clinician's place is not only at the bedside but also in the laboratory, in order to get a comprehensive overview of the patient and his investigations and not try to draw conclusions from either alone. This combined interest in the patient as well as the biopsy has continued throughout my period in Sheffield and latterly at the Hammersmith Hospital.

In our book, *Muscle Biopsy: A Modern Approach*, Dr Michael Brooke and I reviewed the advances in techniques of processing and interpreting the muscle biopsy, against a background of the various clinical syndromes. In this book I have tried to bring together all the clinical syndromes, both old and new, against a background of the muscle biopsy. The two are interdependent and the days are long past when the clinician, however astute, could make a definitive diagnosis on clinical grounds alone, and stand unchallenged.

Any work of this nature is dependent on the help and goodwill of many people; to name them all would entail another chapter to this book. I would like particularly to mention the following: Dr David Lawson at Queen Mary's Hospital for Children for his encouragement and the facilities he placed at my disposal in the early years; Mr Bob Reynolds, Head Physiotherapist at Queen Mary's, who first drew my attention to many of the problems of patients with muscular dystrophy, and Miss Mary Pugh for her excellent photographic documentation of those cases; Professor Peter Daniel at the Institute of Psychiatry, The Maudsley Hospital, who taught me the rudiments of muscle pathology; Professor Everson Pearse at the Hammersmith who provided me with instant bench space and access to the one and only prototype cryostat ('wheezy') then being developed in his laboratory, and fired my interest in histochemistry, and the many colleagues from all corners of the world who converged on his laboratory and were my constant mentors; the late Professor John

Cumings who provided facilities for continuation of this work during my tenure of a lectureship in his department of Clinical Pathology at the National Hospital for Nervous Diseases; Professor Ronald Illingworth for the encouragement and autonomy to pursue my interests during my 11 years at the Children's Hospital in Sheffield; Mr Alan Tunstill for the excellent clinical photography and his continued interest and enthusiasm in this work throughout that period; Mr David Hawtin for the clinical photography at Hammersmith; a succession of superb technicians including Joan Wingfield, Julie Franks (née Binns) and in particular Christine Hutson (née Heinzmann) who has emigrated from her native Yorkshire to London with me; my clinical research associates, Allie Moosa, Mary Cunningham and Gwilym Hosking, and all the short-term research fellows who have been attached to my unit; Charles Galasko, and latterly Sean Hughes, for their interest in the orthopaedic problems; and finally the many paediatric colleagues who referred their patients – without them this book would never have been possible.

Writing a book always seems to start as a challenge and invariably ends up as an intimate love–hate relationship, and I am surely not the first author to be accused by his wife of bigamy. One always owes a particular debt to one's family for their tolerance and understanding during this time of split loyalties.

Throughout the preparation of this book my task has been considerably lightened by the outstanding secretarial expertise of Mrs Valerie Chalk. I would also like to record my thanks to Mr Bill Schmitt and Miss Patricia Terry at W. B. Saunders for their help and enthusiasm.

Much of my research over the years has been generously supported by the Muscular Dystrophy Group of Great Britain. I also wish to acknowledge additional grants from the Medical Research Council, Action Research for the Crippled Child and the Coxen Trust, as well as the munificent grant from the Muscular Dystrophy Association of America in 1975 to establish the Jerry Lewis Muscle Research Centre at Hammersmith Hospital.

VICTOR DUBOWITZ

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Chapter One

DIAGNOSIS

INTRODUCTION

Muscle disorders can be broadly subdivided into *myopathies*, in which the pathology is confined to the muscle itself, with no associated structural abnormality in the peripheral nerve, and the *neuropathies or neurogenic atrophies*, in which the muscle weakness is secondary to an abnormality along the course of the peripheral nerve, from the anterior horn cell to the neuromuscular junction. Both the myopathies and the neuropathies can be further subdivided into hereditary syndromes and acquired ones, and into acute and chronic disorders. In trying to classify the various syndromes it is helpful to follow the anatomical route of the lower motor neurone (Figure 1–1).

Further categorization is based on the characteristic pattern of particular disorders. Thus the term *muscular dystrophy* is used for the genetically determined, progressive, degenerative myopathies, and these are subdivided on the basis of clinical distribution and severity of weakness and mode of inheritance (see Chapter 2, Table 2–1). Similarly, the *spinal muscular atrophies* and *motor neuropathies* are neurogenic disorders in which the lesion is in the anterior horn cell or the peripheral nerve and these are further characterized on the basis of clinical features and mode of inheritance (see Chapter 6).

The so-called *congenital myopathies* are a new generation of genetically determined muscle disorders whose clinical presentation may be somewhat similar to the muscular dystrophies or neurogenic atrophies, but in which specific structural abnormalities can be recognized in the muscle.

The *metabolic myopathies* are akin to the congenital myopathies but represent those syndromes in which a specific metabolic abnormality has been identified, usually related to carbohydrate or lipid metabolism. A further series of syndromes is also included with a presumptive but not yet fully defined metabolic basis.

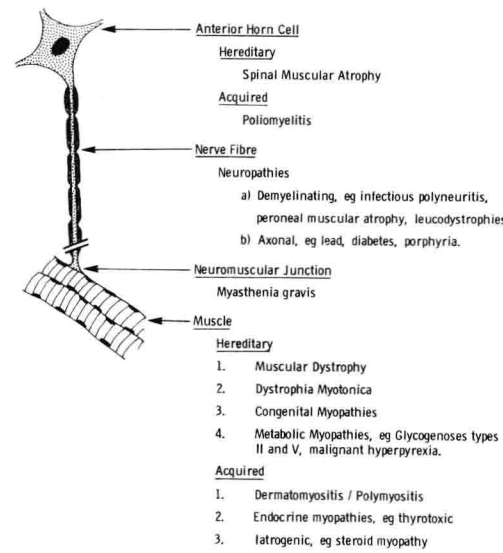

Figure 1–1. Disorders of the lower motor neurone. Anatomical approach. (Courtesy of Dr A. Moosa.)

The *myasthenic* and *myotonic* disorders are recognized on the basis of their specific features, both clinically and on electrodiagnostic investigation.

The various *acquired muscle disorders* include sundry inflammatory, endocrine and toxic conditions affecting either the peripheral nerve or the muscle itself.

The *floppy or hypotonic infant* is a diagnostic problem unto itself and may reflect either a neuromuscular disorder or be associated with a primary disorder in another system, particularly the central nervous system, in which the hypotonia is an incidental feature.

Accurate diagnosis in this wide array of disorders is dependent on a careful clinical assessment followed by the appropriate investigations.

Epidemiology

Muscle disorders are ubiquitous and affect all races. The apparent high incidence of some disorders in particular countries, such as congenital muscular dystrophy in Finland and Japan, may reflect inbreeding in isolated communities increasing the incidence of relatively uncommon recessive disorders.

Statistics on the incidence of various muscle disorders are not readily available and estimates vary widely. Thus the estimated incidence of Duchenne muscular dystrophy, which is probably the commonest neuromuscular disorder in childhood, has ranged from about one in 3000 to one in 8000 live male births, whereas the recent application of a new screening technique produced an incidence of about one in 1700 male births (see Chapter 2, p. 22). The other common neuromuscular disorder in childhood is spinal muscular atrophy, which is inherited as an autosomal recessive character and may well have an incidence approaching other common autosomal recessive disorders such as cystic fibrosis.

CLINICAL ASSESSMENT OF MUSCLE PROBLEMS IN CHILDHOOD

The History

A careful clinical history may often give the clue to the particular disorder that the child has. Common presenting symptoms of muscle disorders in childhood include delay in motor milestones, an abnormal gait, tendency to fall, overt muscle weakness, floppiness or hypotonia, muscle cramps or muscle stiffness.

It is always worth starting with an open question to the parents such as, 'What have you noticed wrong?' and allowing them to give a detailed description of their observations before asking direct questions. This presenting story may at times be so perceptive and precise as to allow a fairly confident diagnosis. Thus recently I was able to diagnose myotonia congenita in a child whose father gave the presenting complaint as, 'It's like this doctor. When we travel on the tube [subway] train between Neasden and Wembley, which is only one stop, she becomes so stiff that she practically stumbles off the train and has to be helped, but after a short while she loosens up and becomes normal again.' Often the description the parents have for a particular gait or the way in which a child manages a particular activity such as getting up from the floor or climbing stairs may give a very good idea of the extent of the child's disability.

In addition to the presenting features one should routinely ask about other associated features suggestive of muscle disorder (see the Questionnaire Appendix at the end of this chapter). It is also worth routinely asking as a direct question whether the particular disability or weakness is getting better, getting worse or remaining static. At times one may get a history of weakness being static over six or 12 months or more in progressive disorders such as Duchenne dystrophy, which highlights the importance of assessing the progression of a chronic muscle disorder over relatively long periods. One may also at times get a history of apparent improvement in Duchenne dystrophy because the child has achieved a new activity, such as riding a tricycle. It is important in such instances to obtain a history as to whether certain specific activities the child had difficulty with, such as climbing stairs, getting up from the floor, running, hopping or skipping, have shown improvement or deterioration.

From the type of problem the child has one can often get an idea as to whether the weakness is proximal or distal or generalized. One should also enquire about any associated difficulty with chewing and swallowing or associated respiratory disorders.

The parents should be questioned as to any variability in the muscle weakness from day to day or from one time of the day to another, or whether there is fatigue on effort and improvement with rest (suggesting myasthenia) or increased disability with inactivity and improvement with activity (suggesting myotonia).

Any change in the appearance of particular muscles, suggesting either an increase in prominence (such as the enlarged calves in Duchenne and other forms of dystrophy) or atrophy, should be noted.

If the patient presents with *cramps* in the muscles one should particularly ask whether these are related to exercise or not. Cramps occurring at rest are usually not of muscle origin and in most cases do not warrant any further neuromuscular investigations. These are the commonest cramps occurring in childhood. Cramps related to exercise and relieved by rest usually point to a muscle disorder (see Table 1–1). They are a presenting feature of some of the more benign and chronic

Table 1–1. Causes of Muscle Cramps

A. Occurring at rest. Usually not muscle disorder.

B. Occurring with exertion. Relieved by rest. May be associated myoglobinuria.

1. Benign and low-grade muscular dystrophy (limb girdle or Becker types)

2. Metabolic disorders
   (a) Glycogenoses:
   type V (phosphorylase deficiency)
   type VII (phosphofructokinase deficiency)
   (b) Lipid metabolism disorders:
   carnitine palmityl transferase deficiency

3. Other myoglobinuric syndromes

Table 1–2. Causes of Episodic Weakness

1. Periodic paralysis
2. Relapsing polyneuropathy
3. Dermatomyositis; polymyositis
4. Myasthenia gravis
5. Rhabdomyolysis

forms of muscular dystrophy such as the Becker and limb girdle varieties (see Chapter 2). There is usually associated weakness. Cramps on exertion are also a presenting feature in some of the rare metabolic myopathies such as the glycogenoses associated with phosphorylase and phosphofructokinase deficiency and the disorders of lipid metabolism associated with deficiency of the enzyme carnitine palmityl transferase. An enquiry of possible associated myoglobinuria should be made in these cases. The muscle power of these patients may be normal. Muscle cramps are also an associated feature of other myoglobinuric syndromes (see Chapter 4).

If the patient presents with recurrent episodes of muscle weakness one should think of one of the syndromes with periodic weakness (Table 1–2) and investigate appropriately.

Family History

A detailed family history and pedigree chart is essential in every case in order to establish a possible genetic mechanism. In X-linked conditions one should get as much detail as possible particularly of male relatives on the maternal side and it is also worth noting female relatives who may be available for investigation to establish possible carrier status.

In autosomal recessive conditions the earlier generations may be normal and the parents of affected siblings are presumptive heterozygotes. Consanguinity of the parents will increase the chances of carrying the same genes.

In dominantly inherited conditions such as dystrophia myotonica and facioscapulohumeral dystrophy there is often marked clinical variability within a family and careful examination of apparently symptom-free relatives may reveal affected cases.

Before giving genetic counselling in any family it is essential first to establish as accurately as possible the diagnosis in the index case.

Clinical Examination

One can often acquire a lot more information on the degree and localization of *muscle weakness* in a small child by observing some of his spontaneous activities before trying to do a formal assessment along the lines of the traditional 'adult' neurological examination. Attempts to undress and forcibly restrain a child in a supine position on an examination couch are likely to end up with a thoroughly uncooperative and irritable child and an equally irritable and frustrated examiner.

The child should be carefully observed from the time he comes into the consulting room and throughout the period that the history is being obtained from the parents. His gait should be noted and his posture in the standing position. If he is not ambulant, note his general activity or lack of it. The face as well as the limbs should be observed.

A number of assessments can be done while the child is still clothed. These include: a detailed assessment of his gait, whether he walks on a wide or narrow base, on his toes or on the flat feet, whether the feet are excessively everted, and whether he can walk on his heels or along a straight line heel to toe; his ability to get up from a chair, to sit up from the supine, to go down on to and to get up from the floor, to go up and down two or three steps, to jump on both legs, to stand on one leg and to hop on one leg.

A small infant can be engaged in play activity and given small cubes to handle and to place on top of each other. This will help assess his co-ordination as well as his ability to raise the arms against gravity.

These general assessments will help to decide if there is any obvious weakness present and also whether its location is proximal, distal or generalized. The child who is able to hop on one leg and to get up from the floor without any difficulty and without supporting a hand on the knee is unlikely

to have any significant weakness in the lower limbs. The examination can then be followed by a more detailed assessment of individual muscle groups after the child is undressed.

Table 1–3. Some Motor Milestones in Normal Development (Average)

Birth:	Flexor posture of limbs, able to sustain head in line with body in ventral suspension and supine.
3 months:	Momentary sitting posture, flexed forward.
6 months:	Sits leaning forward on hands. Lifts head spontaneously in supine. Takes weight on legs well.
9 months:	Sits. Pivots. Pulls to standing position.
1 year:	Walks with help. Cruises.
15 months:	Walks unaided. Creeps up stairs.
18 months:	Walks without falling. Runs. Climbs stairs unaided.
2 years:	Able to run well. Kicks a ball. Goes up and down stairs.
3 years:	Stands on one leg. Jumps off a step.
4 years:	Hops on one foot (just).
5 years:	Good ability to hop on either leg.

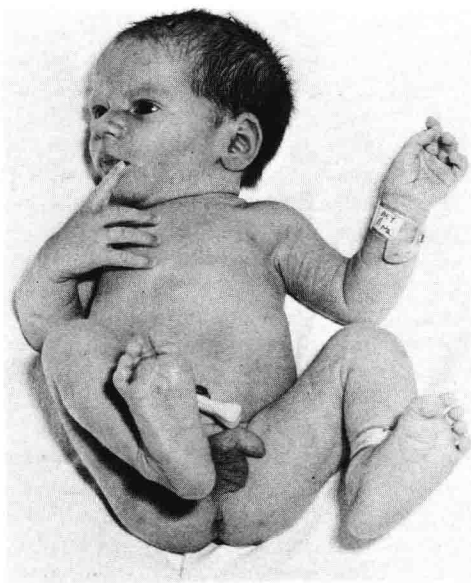

Figure 1–2. Newborn infant showing flexed posture of limbs and ability to maintain arms and legs against gravity. Note expressive face and closed mouth.

In each age group there are milestones of motor development which are helpful in assessing the integrity of the neuromuscular system. The clinical examination should thus be geared to the age of the child (Table 1–3).

In the neonatal period and in very young infants one can observe general mobility of face and limbs (Figure 1–2). If the limbs are not being moved spontaneously one can observe the response to a stimulus such as stroking the soles, or the ability to sustain a passively elevated arm or leg. The control of the head and limbs in ventral suspension and while supine with traction on the hands will give a good general idea of abilities and disabilities. Even in the immediate newborn period a full-term infant should be able to keep the head in line with the body in both the prone and supine postures and should also have a good degree of flexor tone in the limbs in ventral suspension (Figures 1–3 and 1–4). These manœuvres can be

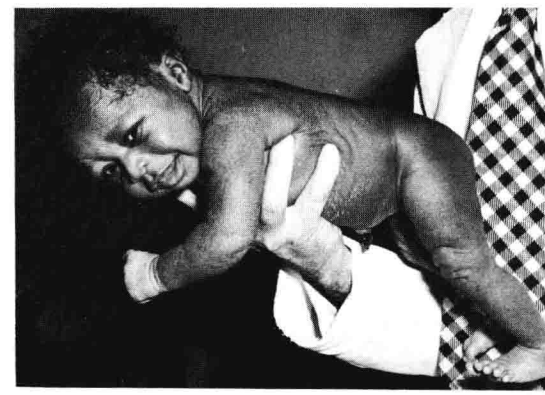

Figure 1–3. Newborn infant. Ventral suspension showing ability to maintain head in line with body. Note flexed posture of limbs.

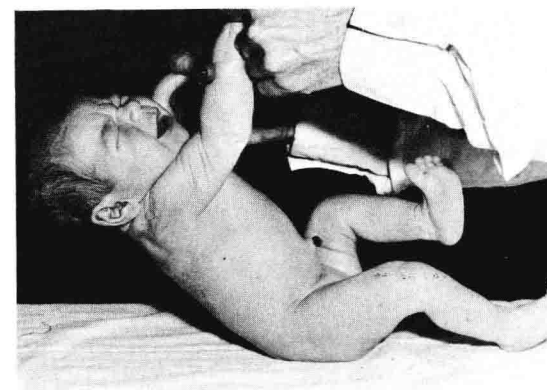

Figure 1–4. Newborn infant showing ability to maintain head in line with body in supine posture with traction on hands. Note sustained elevation of leg against gravity.

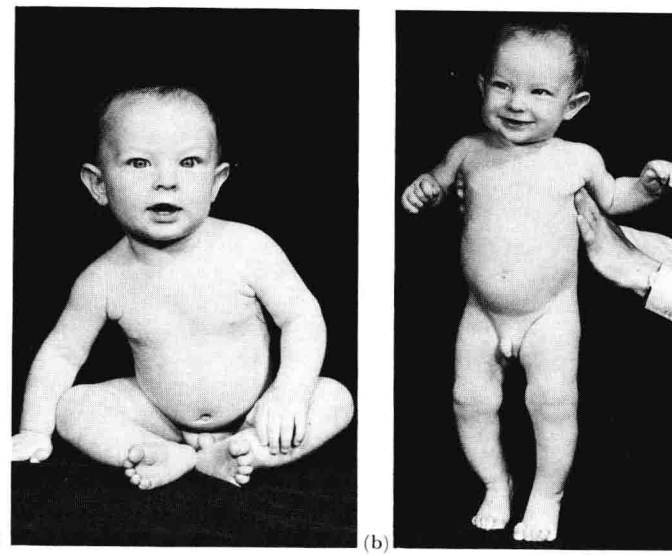

Figure 1–5. Seven-month-old infant showing ability (a) to sit steadily without support and (b) to sustain weight of body on legs.

Figure 1–6. 'Parachute' response, showing outstretching of arms when held in a head-down position.

Figure 1–7. 'Wheelbarrow' posture. Ability to take weight of body and locomote with hands. (Same infant as Figure 1–6.)

used throughout the first three months, and one should again look for the degree of head control and the mobility of the limbs.

From the age of three months onwards one can also note the child's ability to reach out for objects, his ability to support weight on the legs when held vertically and bounced on his feet, and his head control in the vertical, prone and supine positions.

Later milestones such as sitting without support, pivoting, and standing with and without support are useful (Figure 1–5). In addition to observing the weight-bearing ability of the legs, one can also assess the mobility of the arms in the 'parachute' response when the child pushes out his hands when he is inverted and moved head-first towards the floor (Figure 1–6). This is usually well developed by about nine months of age. The power in the arms and shoulder girdle muscles can be assessed in the 'wheelbarrow' posture by his being kept suspended with his head down and made to 'walk' on his hands (Figure 1–7).

Once he starts walking one can observe his ability to get up from a sitting, prone or supine position on the floor, and his ability to go up and down steps (a simple two-step wooden construction with non-slip rubber sheeting on the steps is useful for outpatient purposes).

After the age of three years he should be able to stand on one leg; from the age of four to jump on both legs and after the age of five to hop on one leg. From the age of three onwards one can usually get the co-operation of the child in attempting various manœuvres.

In assessing the shoulder girdle one should also note, in addition to any weakness and difficulty

with movements, the upward and forward riding of the scapulae with abduction of the arms, giving a very typical appearance in facioscapulohumeral syndromes (see Chapter 2, Figure 2–49).

The face can be assessed by noting its appearance at rest, the presence of an open drooping or tent-shaped mouth, and whether there is lack of movement and expression. The power can be assessed by getting the child to screw his eyes tightly closed – normally he should be able to bury his eyelashes completely (Figure 1–8). In the presence of weakness, even of mild degree, the eyelashes will remain out. The lower face can be assessed by getting the child to show his teeth, smile, puff out his cheeks and pout his lips, and to blow or whistle.

Once a general appraisal has been made in this way one can go on to more detailed quantitative assessment of individual muscle groups. The standard Medical Research Council (MRC) classification (1943) provides a useful baseline (Table 1–4). This designates five categories ranging from zero movement in the muscle (0) to full strength (5) and antigravity power in between (3). Grade 1 denotes a flicker of movement only, grade 2 contraction short of antigravity power, and grade 4 power more than antigravity but not full.

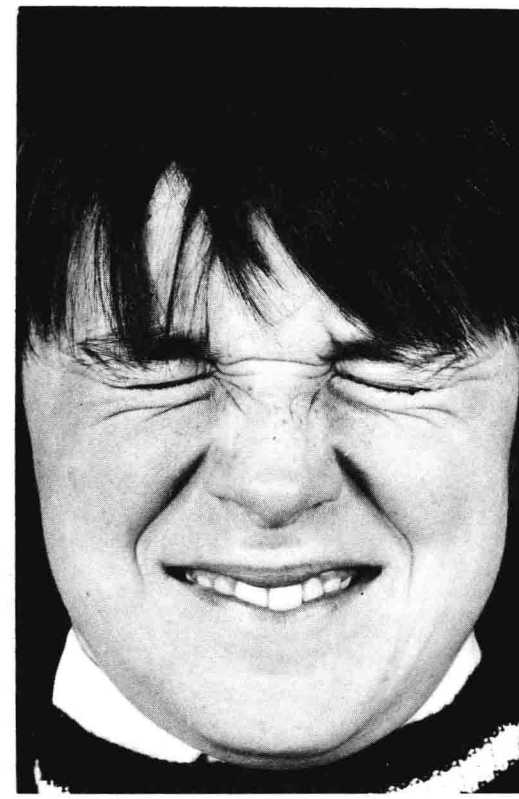

Figure 1–8. Ability to screw up eyes tightly and bury the eyelashes completely. Note also the symmetrical facial movement with showing of teeth.

Table 1–4. MRC Scale for Evaluation of Muscle Power

0 – No contraction
1 – Flicker or trace of contraction
2 – Active movement, with gravity eliminated
3 – Active movement against gravity
4 – Active movement against gravity and resistance
5 – Normal power

Table 1–5. Minimum duration for holding head or leg in 45° of flexion in normal supine child at various ages

Minimum time head held up (seconds)			
Children aged	5–6 yr	(n=47)	12
	7–10 yr	(n=170)	30
	11–15 yr	(n=78)	60
Minimum time leg raised to 45° in supine (seconds)			
Children aged	5–6 yr	(n=45)	60
	7–10 yr	(n=169)	40
	11–15 yr	(n=92)	80

n = number.

As a screening test for possible muscle weakness we have found it useful to time the child's ability to sustain the neck or straight leg in a position of 45° of flexion while lying in the supine position (Hosking et al, 1976). Although there is a very wide range for the duration of this manœuvre in normal children at different ages, we have found that the lower limit of our normal range (Table 1–5) provides a useful cut-off point, and no children of equal age with Duchenne dystrophy could maintain elevation of the neck or leg for this duration.

The recent development of a sensitive handheld myometer (Figure 1–9) by Edwards and McDonnell (1974) has made possible a more objective measurement of the actual force exerted by a particular group of muscles. We have found this myometer to be practical in the childhood period as well, although different grades of force and calibration are necessary in weaker cases (Hosking et al, 1976). It is of particular value in assessing the progress of weak muscles but can also provide a useful index of the degree of weakness in a particular muscle in comparison with