

POSTURE IN MUSCULAR DYSTROPHY - THE COMPENSATORY ROLE OF PSEUDOHYPERTROPHY, EQUINUS AND LORDOSIS

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ABSTRACT

THIS PAPER REPORTS ON A BIOMECHANICAL ANALYSIS OF THE CHARACTERISTIC POSTURE OF MUSCULAR DYSTROPHY. IT IS POSTULATED THAT THE MUSCULAR DYSTROPHY CHILD ADOPTS LORDOSIS AND EQUINUS TO COMPENSATE FOR WEAKNESS IN THE HIP AND KNEE EXTENSORS AND FOR KNEE FLEXION CONTRACTURES. STUDIES WERE CARRIED OUT ON NORMAL SUBJECTS WHO SIMULATED THE POSTURE OF MUSCULAR DYSTROPHY AND ON DYSTROPHIC CHILDREN UTILIZING A FORCE PLACE, CINEPHOTOGRAPHY, RADIOLOGICAL METHODS AND E.M.G. THE RESULTS VERIFY THAT THE TYPICAL POSTURAL ALTERATIONS IN MUSCULAR DYSTROPHY EFFECTIVELY STABILIZES THE KNEES.

INTRODUCTION

PROGRESSIVE MUSCULAR DYSTROPHY IS A GENETICALLY DETERMINED DEGENERATIVE PROCESS OF SKELETAL MUSCLE FIBRES. THE DISEASE TAKES A VARIETY OF CLINICAL FORMS RESULTING IN WEAKNESS WHICH PROGRESSES TO TOTAL IMMOBILITY. IN THE CHILDHOOD FORM, A BIZARRE POSTURE DEVELOPS WITH CHARACTERISTIC LORDOSIS EQUINUS AND ENLARGEMENT OF THE CALF MUSCLES SO CALLED "PSEUDOHYPERTROPHY". NO UNIFIED CONCEPT EXISTS TO EXPLAIN THE COMPONENTS OF THE POSTURE. LORDOSIS IS CLASSICALLY ATTRIBUTED TO WEAK ABDOMINAL LOW BACK AND HIP EXTENSOR MUSCLES, AND FLEXION CONTRACTURES OF THE HIPS AND THE CALF PSEUDO - HYPERTROPHY TO FATTY INFILTRATION. A NUMBER OF AUTHORITIES (1,2) SUGGEST THAT THE CALF ENLARGEMENT REPRESENTS TRUE HYPERTROPHY BUT ARE AT A LOSS TO EXPLAIN WHY IT DEVELOPS.

MILLER (3) HAS MADE THE FOLLOWING CLINICAL OBSERVATIONS:-

- 1) CHILDREN WHO WALK FAIRLY WELL WITH EQUINUS CONTRACTURES ARE TOTALLY DISABLED BY HEEL CORD LENGTHENING.
- 2) APPLICATION OF KNEE BRACES TO THOSE CHILDREN RESTORES AMBULATION AND ALSO REDUCES THE LORDOSIS TO A CONSIDERABLE DEGREE.
- 3) HIP FLEXION CONTRACTURES IN THE AMBULATORY PATIENT ARE TOO SMALL TO EXPLAIN THE LORDOSIS.
- 4) IN CONTRAST TO PROFOUND WEAKENING OF HIP AND KNEE EXTENSOR, THE CALF MUSCLES RETAIN A LARGE PROPORTION OF THEIR STRENGTH AND BECOME THE STRONGEST MUSCLES IN THE LOWER LIMBS.
- 5) EQUINUS OCCURS "ELECTIVELY" IN SOME PATIENTS WITHOUT FIXED DEFORMITY AT THE ANKLE WHEN THEY ARE ON UNEVEN FOOTING OR WHEN APPREHENSIVE.

ON THE BASIS OF THESE OBSERVATIONS, IT IS POSTULATED THAT THE POSTURAL CHANGES ARE IN PART COMPENSATORY. THE EQUINUS SHIFTS THE CENTRE OF

FOOT PRESSURE TOWARD THE METATARSAL HEADS. EQUINUS AND LORDOSIS ADVANCE THE CENTRE OF GRAVITY OF THE BODY RELATIVE TO THE KNEE AXIS. THE RESULT WOULD BE STABILIZATION OF THE KNEE IN EXTENSION EVEN IN THE PRESENCE OF WEAK KNEE EXTENSORS AND KNEE FLEXION CONTRACTURES.

EXPERIMENTAL STUDIES:

TO DETERMINE THE STABILIZING MOMENT AT THE KNEE JOINT, A SERIES OF EXPERIMENTS WERE PERFORMED ON (A) NORMAL ADULTS AND CHILDREN SIMULATING THE LORDOSIS AND EQUINUS OF MUSCULAR DYSTROPHY AND, (B) TWO CHILDREN WITH THE DUCHENNE FORM OF MUSCULAR DYSTROPHY.

A FORCE PLATE (4) WAS USED TO RECORD THE REACTIVE GROUND FORCES AND ITS COORDINATES. THE ORIGIN OF THE COORDINATE SYSTEM WAS LOCATED AT THE FIFTH METATARSO-PHALANGEAL JOINTS. SIMULTANEOUS 16 MM. CINE RECORDS OF THE SUBJECTS' LOWER LIMB SEGMENTS WERE TAKEN IN THE CORONAL PLANE AGAINST A BACKGROUND GRID. THE POSITION OF THE AXIS OF ROTATION OF THE KNEE AND ANKLE JOINTS (5,6) WERE IDENTIFIED BY X-RAYS AND SKIN MARKERS. THE REACTIVE GROUND FORCE AND ITS COORDINATES, E.M.G. RECORDS OF SOLEUS AND GASTROCNEMIUS ACTIVITY, AND A SYNCHRONIZATION PULSE FROM THE MOVIE CAMERA WERE RECORDED ON A 7 CHANNEL MAGNETIC TAPE RECORDER, AND SUBSEQUENTLY REPRODUCED IN STRIP CHART FORM. THE E.M.G. RECORDS WERE FULL WAVE RECTIFIED AND FILTERED TO QUANTITATE MUSCLE ACTIVITY. SINGLE FRAME ANALYSIS OF THE CINE FILM DETERMINED COORDINATES OF THE AXIS OF ROTATION.

DATA WAS TRANSFERRED TO PUNCH CARDS AND CALCULATIONS PERFORMED ON A DIGITAL COMPUTER.

CALCULATION OF THE PLANTAR FLEXING TORQUE AND MOMENTS AT THE KNEE JOINT WERE BASED ON THE LEG MODEL IN FIG. 1.

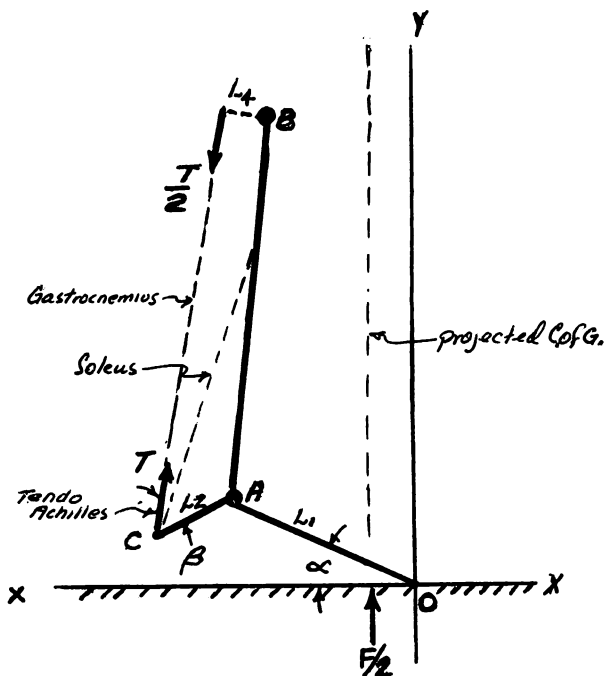


FIG. 1

THE TENSION IN THE ACHILLES TENDON WAS CALCULATED BY SUMMATION OF MOMENTS ABOUT THE ANKLE AXIS A. THE NET STABILIZING MOMENT (N.S.M.) AT THE KNEE JOINT WAS CALCULATED BY SUMMATION OF THE DESTABILIZING MOMENT DUE TO TENSION IN THE GASTROCNEMIUS MUSCLE WHICH HAS ORIGIN ABOVE THE KNEE JOINT, AND THE STABILIZING MOMENT DUE TO THE GRAVITATIONAL FORCE ACTING ON THE BODY MASS ⁽⁷⁾ ABOVE THE KNEE JOINT. THE VALUE OF THE TENSION EXERTED BY THE GASTROCNEMIUS USED IN CALCULATING THE DESTABILIZING MOMENT AT THE KNEE WAS 50% OF THE TENSION (T) IN THE ACHILLES TENDON. THIS PERCENTAGE WAS DETERMINED ON THE BASIS OF RELATIVE MUSCLE WEIGHTS AND BY EVALUATION OF E.M.G. RECORDS. THE FORCES DUE TO OTHER MUSCLES ACTING ACROSS THE KNEE AND ANKLE JOINTS CAN BE CONSIDERED INSIGNIFICANT ON THE BASIS OF E.M.G. STUDIES OF SYMMETRICAL STANCE. THE PASSIVE TISSUE FORCES AT THE ANKLE WERE IGNORED AND IN DOING SO A CONSTANT WAS INTRODUCED WHICH INCREMENTED THE TENSION IN THE ACHILLES TENDON. THIS BIASES THE NET STABILIZING MOMENT TO A LOWER VALUE, BUT ABSOLUTE DIFFERENCES IN VARIOUS STANCE MODES ARE UNAFFECTED.

RESULTS

IN STUDIES OF NORMAL INDIVIDUALS SIMULATING DYSTROPHIC POSTURE AN INCREASE IN THE N.S.M.* DUE TO LORDOSIS ALONE WAS 90% IN THE ADULT AND 108% IN THE CHILD. EQUINUS WITHOUT LORDOSIS INCREASED THE N.S.M. MAXIMALLY AT 8 DEGREES OF EQUINUS, BY 52% IN THE ADULT AND 45% IN THE CHILD. EQUINUS WITH LORDOSIS INCREASED THE N.S.M. BY 262% IN THE ADULT AND 280% IN THE CHILD, AGAIN AT 8 DEGREES OF EQUINUS.

THE RESULTS OF STUDIES OF THE DYSTROPHIC

*NET STABILIZING MOMENT AT THE KNEE JOINT

CHILDREN REVEALED AN N.S.M. IN PLANTIGRADE STANCE OF LESS THAN 50% OF THE VALUE TO BE EXPECTED WHEN WEIGHT AND HEIGHT CORRECTION WAS APPLIED. WHEN THE SUBJECTS INTRODUCED FURTHER EQUINUS THEY ACHIEVED LESS THAN 10% OF THE INCREASE IN THE N.S.M. OBTAINED IN A NORMAL SUBJECT OF SIMILAR HEIGHT AND WEIGHT. THE MEAN VALUES OF THE HEEL CORD TENSION IN PLANTIGRADE STANCE WERE ALSO FOUND TO BE INCREASED WHEN COMPARED TO NORMAL SUBJECTS.

CONCLUSIONS

THESE STUDIES SUPPORT THE CONTENTION THAT THE TYPICAL POSTURAL ALTERATIONS IN MUSCULAR DYSTROPHY (LORDOSIS AND EQUINUS) HAVE A STABILIZING EFFECT AT THE KNEE. IN CONJUNCTION WITH CLINICAL OBSERVATIONS, THEIR COMPENSATORY ROLE IS GIVEN FURTHER SUPPORT. THE FAILURE OF THE DYSTROPHIC CHILDREN TO INCREASE THE N.S.M. TO AS GREAT A DEGREE AS THE NORMAL SUBJECT WHEN ADOPTING EQUINUS CAN BE EXPLAINED BY THE FACT THAT BOTH CHILDREN HAD SMALL KNEE FLEXION CONTRACTURES, LORDOTIC COMPENSATION AND UNMEASURABLE EQUINUS HAD ALREADY SHIFTED THE REACTION FORCE CLOSER TO THE TOES. THE HIGH TENSION FORCES OCCURRING IN THE ACHILLES TENDON OF THE DYSTROPHIC CHILDREN SUPPORTS THE CONCEPT THAT THE ENLARGED CALF MUSCLES DO UNDERGO A TRUE WORK HYPERTROPHY.

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