MULTIPLE CONGENITAL ARTICULAR RIGIDITIES

A Review of the Literature with Reports of Two Cases II.

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Part II

Abstracts of Cases

Otto, A. 1841.—Described condition as "Monstrum humanum trunco nimis brevi et extremitatibus incurvatis." Female fœtus age 7 or 8 months. Very short neck. Stiff and short trunk. Arms close to sides and elbows flexed by contracted muscles. Thighs flexed on abdomen and legs on thighs. Hands and feet deformed, feet in equino-varus. Head large, mouth small with complete cleft-palate. Chest, lungs and heart small. Thymus, large. Abdominal viscera in normal position but kidneys low at the brim of the pelvis. Spinal column normal as were the bones of the extremities.

Baumgartner 1890 (cit. Wunsch).—Flexion contracture of elbows. Posterior subluxation of ulna. Volar and ulnar flexed club-hands. Flexion of fingers with webbed bases. Hips in flexion. Right pes equinus, left pes equino-varus.

Wirt, W. E. 1891.—Contracted shoulders and elbows with much diminished movement. Flexed hands and fingers. Contracted hips with a right dislocation. Flexed knees and rudimentary condyles and patellæ, latter displaced laterally. Movement of knees produced crackling in the joints. Internal ligament of knee very lax making lateral movement possible. Bilateral pes equino-varus.

Redard, P. 1893.—Female, age 2 months. Family history negative. Parents healthy but paternal aunt an idiot. Five robust siblings. Uneventful pregnancy. Difficult birth. Birth fracture of humerus. Hips fully flexed, knees extended, clubbed feet on front of chest at root of neck. Great toes, flexed. Absent patellæ. Upper limbs extended at sides of trunk. Wrists, hands, fingers flexed. Principal muscles of upper limbs, hard, rigid and incapable of active or passive movement. "*Il ressemble á une poupée en bois sans articulations.*" All movements fingers and wrist impossible. No flexion of elbows. Difficulty in palpating joint outlines. All shoulder movement abolished. Striking absence of articular creases and uniform cylindroid appearance of limbs. At 2½ years a bright child, no defect of special senses. Contractures did not disappear under chloroform anæsthesia. No reaction of degeneration only extensors of forearm did not respond. Much intra-articular crackling on forced flexion of elbow joints, indicating rupture of fibro-tendinous parts producing the peri-articular resistance.

Case 2.—Male, aged 7½ years. Parents well. Three normal siblings. Pregnancy normal. Normal presentation, rapid birth. Thighs flexed on pelvis, and close to abdomen. Knees flexed. Feet twisted backwards and clubbed. Toes flexed. Patellæ absent. Lower limbs adopted diamond

shape. Extension of knees impossible but some flexion. Thighs abducted, rotated forwards, limited adduction and posterior rotation (passive). No voluntary movements of lower limbs and in these circulation extremely poor. Electrical reactions and physiological reflexes, normal as were special senses and intelligence.

CASE 3.—Female, 10 years old. Healthy parents. Difficult labour. Abnormal presentation. Left lower extremity marked abduction. Leg flexed on thigh. Foot in equino-varus. Flexion and abduction of hip normal. Extension and adduction slightly limited. Crackling in knee joint with forced passive movement. No active movement of knee joint. Patellæ rudimentary, just a fibrous nodule. Toes " en griffe." Poor circulation in skin. Excess of subcutaneous tissue. Atrophy affecting whole of left lower extremity, especially the leg. Reflexes diminished, electrical responses normal.

BRODHURST (cit. Redard).—Flexed upper extremities, extended lower. Contractures of feet, knees and hips.

ADAMS (cit. Redard).—Double club-foot. Flexed knees, atrophied patellæ. Fingers and hands fixed rigidly in extension.

BOUVIER (cit. Redard).—Fœtus of 7 months. Double club-foot. Right club-hand. Extended knees, flexed thighs. Flexed left elbow, extended right elbow.

SCHANZ, A. 1898.—Male, age 4 years. Movement of mandible limited to 2 cm. and sideways movement in one direction only. Neck short and movements limited. Hypertrophied platysma muscle. Thoracic and lumbar spine also limited to a degree. There were contractures about the vertebral column and especially about the atlanto-occipital joint. Extremities tapered distally. Shoulders high and movement restricted. Active abduction 90°, passive 120°, forward to 45° above the horizontal. Supination, not quite full, as were the hand movements. Fingers semi-flexed and stretching produced tightening of subcutaneous tissue, with backspringing of fingers which were webbed at the bases, including thumb. Fifth finger could not be included in fist. Hips in abduction, knees in semi-flexion, right more than left, they flexed to 100° and extended 20°. " Webbing " of popliteal tissue on extension. Patellæ subluxated laterally. Knees in marked genu valgum position. Severe right club-foot, left flat-foot, formed a " compression unit."

WOLF, J. (cit. Schanz).—Age $1\frac{1}{4}$ years. Contracture deformities in extension of all extremities. Little movement of shoulders. Elbows rigid, in extension, also knees. Thighs abducted and externally rotated. Bilateral pes equino-varus and club-hand.

WUNSCH, M. 1901.—Male, age $3\frac{1}{2}$ years. Some asymmetry of skull. Right elbow extended to 135°, flexed to 70°, both actively and passively. Left elbow, normal. Pronation and supination, good. Thumbs extended, fingers slightly flexed. Some dorsi-flexion of hand possible. Ulnar styloid not palpable. Interphalangeal contractures of fingers. Complete flexion of knees possible, but right extended to 100° only, left to 135°. Bilateral club-foot.

VON ZENGERLY (cit. Wunsch).—Atrophied upper extremities, but elbows and shoulders free. Hands in volar and ulnar flexion and fingers in flexion. Atrophied lower extremities, hips free, also right knee. Left genu-valgum. Bilateral pes equino-varus. Electrical reactions normal.

BEDNAR (cit. Wunsch.).—Contractures of elbow-joints and club-hands. Contractures of knees and club-feet. Some scoliosis.

Phocas, M., 1899.—Male, age 3½ months. Parents healthy. Only child. When mother 6 months' pregnant fell and suffered abdominal pains. Birth normal at term. Head somewhat large. Knees flexed 125°, with 5° extension. Abduction of thigh impossible at first. Patellæ slightly displaced in front of femoral condyle. Subluxation of tibia. Contracted popliteal tendons. Testes in inguinal canals.

Case 2.—Male, age 2½ years. Parents healthy. Flexed knees on which patient walked (very few details).

Case 3.—Female, age 1 month. Healthy parents. Born with flexed knees. Right knee extended to 140°, left to 177°.

Case 4.—Male, age 3½ years. Parents well. Only child. Difficult pregnancy—mother ill during last 2 months. Patient born with flexed hips and knees. Bilateral pes valgus. Difficult to separate the limbs. Patellæ present. Skin dimples over lateral aspect knees. Knees could not be completely extended but with maximum extension there was a genu valgum. Feet flat and in valgus.

Case 5.—Male, 5 years old. Parents well. Difficult pregnancy, mother ill throughout. Normal birth. Left hand in flexion, right in extension. Feet in talo-valgus. Knees flexed 115°. Femoral condyles large. Tendons cord-like in popliteal fossæ. Marked atrophy of muscle. Cryptorchidism.

Case 6.—Male age 10 months. First-born. Parents well. Patient's general condition poor. Four extremities flexed. Upper limbs at sides, fully flexed elbows. Fingers flexed, thumbs in palms. Thigh flexed on pelvis, leg on thigh, feet in equino-varus position. Great toes flexed, other toes extended.

Phocas, M., and Paucot, 1901.—Male, age 6 years. Father and grandfather alcoholic. Four siblings, one with an infirmity of upper limbs. Mother had serious fall in 6th month of pregnancy. Full term. Left knee flexed 120°—the limit of extension. Patella located with difficulty. External condyle seemed larger than normal. Tendons in popliteal fossa prominent. Slight external convexity of tibia. Flat-foot with some valgus. Atrophy of muscles of thigh and leg. Child walked on his knees which had developed subcutaneous bursæ. Under anæsthesia no change in degree of flexion and tenotomy of popliteal tendon made little difference.

Case 2.—Female, age 2 years. Parents well. No familial diseases. Five siblings, living and well. Abnormal pregnancy—menstruated first trimester, dysmenorrhœa. Fall in 4th month followed by abdominal pain, confined to bed for several days. Two months later fell again—injured abdomen—again confined to bed with severe abdominal pains. Normal delivery. Post-partum hæmorrhage. Patient born with arms folded against chest, knees and hips acutely flexed. Anonychia of toes but nails grew at 4½ months. Upper half of face ill-formed—head large—orbital sockets empty. Arms short compared with forearms—elbows flexed—flexion could be increased but not extension. Easy pronation and supination. Biceps no more than a thin tense cord. X-ray showed deformity of distal articular surface of humerus. Abnormal incurving of radius with subluxation on ulna. Sigmoid surface of radius rather distant from articular surface of humerus.

Inferior Extremities.—Normal hip movement. Thighs short compared with legs. Knees flexed to right angle, could be further flexed but not extended. Aplasia anterior aspect knee and absent patellæ. Posterior subluxation of tibia on femur. Legs also internally rotated on thigh with lateral convexity.

Bilateral flat-foot with slight varus. No change under anæsthesia showed that it was not muscular tissue, but the osseous and articular structures which maintained the deformity and prevented operative correction.

MAGNUS, F., 1903.—Male, age 15 years. Seven sisters, all normal except 1 died of unknown cause. No familial history of deformity. Normal mental and psychological development for age. Brachycephalic. Shoulders high, clavicles short. Scapulæ small, outline well seen under skin. Arms short, very thin, as though only bone covered with skin. Triceps capable of producing forceful extension of elbow. Flexion of elbow limited to 90°. No active pronation or supination. Fingers contracted "en griffe." Arms adducted, internally rotated, hands crossed in front. Atrophic interossei muscles. Sat in oriental position, flexed knees and hips. Marked pes equino-varus, bilateral. Crackling in several joints on passive movement. Attempted extension of knees produced webbing of popliteal space. Patellæ impalpable. Quadriceps muscle not apparent. Abnormality of knee-joint shown by roentgen study. Patellæ not visible on film. On resection of joint, patella, 5 mm. by 2 cm. by 2·5 cm. found between condyles.

SCHMIDT, G., 1904.—Male, age 7 weeks. First child. Much amniotic fluid. Parents cousins. No familial disease. Knees flexed, hips flexed in external rotation and abduction. Hips extended to 90° and adducted to mid-line. Internal rotation greatly limited. Knees flexed fully, right extended 110°, left 90°, tendons became prominent in popliteal fossæ. Marked bilateral pes equino-varus.

ROSENKRANZ, E., 1905.—Male, age 10 years. Three normal siblings, good family history. Normal pregnancy. Normal amniotic fluid. Had already received treatment from first year of life. Intelligence good. Both arms weaker and shorter than normal. Left shoulder muscle more atrophied than right. Flexors of left elbow not palpable. Right biceps tendon just palpable with effort of flexion. Shoulder free. Complete flexion of elbows not possible. No active flexion left elbow, slight of right. Hands in moderate flexion and ulnar deviation. Right forearm midway between supination and pronation, left in full pronation. Fingers in slight flexion, right middle finger worst. Webbing at bases. Thumbs adducted and opposed, active only in this direction. Tips of thumbs and fifth finger atrophic on right. Electrical reactions showed a distally shifting point of stimulus for the muscles supplied by radial nerve except m. ext. carpi ulnaris, m. ext. poll, brevis and ulnar extensors of fingers. No reaction of degeneration.

LUMSDALE (cit. Rosenkranz).—Bilateral extension contracture of fingers. Bilateral club-foot. Flexed knees, rudimentary patellæ.

CONRAD (cit. Rosenkranz).—Left club-hand. Bilateral club-foot. Bilateral flexion contracture of knees and hips. Little amniotic fluid at birth.

CRUVEILHIER (cit. Rosenkranz).—Bilateral club-hand. Extended elbows. Flexed hips, extended knees.

HERMANN (cit. Rosenkranz).—Bilateral club-hand, extended elbows, rigid shoulders. Little flexion in knees, left pes varus.

BEELY (cit. Rosenkranz).—Bilateral club-hand with flexed fingers. Thumbs in palms. Flexed elbows, fixed shoulders. Bilateral club-foot, extended knees, impalpable patellæ, flexed and externally rotated hips. Asymmetric head, paralysis of left face.

MENCIERE (cit. Rosenkranz).—Right club-hand. Flexed right elbow. Right pes equino-varus. Genu valgum. Flexed right knee.

VERNEUIL (cit. Rosenkranz).—Bilateral club-hand, flexed fingers, hands atrophic, thumbs adducted. Movements of elbows and shoulders almost absent.

HOWARD, R., 1908.—*Case, one of twins born at term.* Vertex presentation. Parents healthy. Normal pregnancy. Other twin stillborn (with slight deformity of feet). Case lived 7 days. Body a little shapeless and flattened from side to side. Buttocks flattened, anal cleft almost absent. Anus conspicuous, perineum prominent. Scrotum and penis rather flattened.

Upper extremities.—Arms plump, not well-formed. Six inches long, cylindrical. Roundness of shoulders absent. Arms internally rotated. Olecranons looking forward. Forearms half-supinated. Elbows and wrists flexed. Hands, deviated medially, thumbs slightly abducted. Attempted correction resulted in limbs springing back to original position.

Lower Extremities.—Not well developed. Normal amount of subcutaneous tissue. Thighs flexed on body to little more than a right angle. Legs at right-angles to thighs. Feet in marked talipes equino-varus. Small depressed areas of skin on outer aspect of knees. Straightening of feet resulted in backspringing.

CASE 2.—A case of Hutchinson, with upper limbs in position of Erb's palsy and with similar amount of function as in Howard's case.

CASE 3.—Child 9 years old. Bilateral talipes equino-varus. Both upper extremities in position of Erb's palsy, the only active muscles being those of trunk, head, and neck.

SCHLIVER, K. 1910.—Female, 2½ years old. Family history negative. Parents healthy. Normal sibling. First child. Forceps delivery. Did not move arms until 6 months old, but moved legs from birth. After second birthday stood with support.

Upper Extremities.—In position of Erb's palsy. Shoulders fairly well developed. Soft tissues of arm, flabby. Forearms well developed. Free passive movement of shoulders—active movement limited to angle of 90° on right, a little more on left. Active and passive movement of right elbow limited to 30°, left to 60°. Pronation and supination good. Wrists, hands, fingers, normal.

Lower Extremities.—Soft and flabby muscles. Normal passive movement of hips. Flexion of knees, only 45°, with much lateral mobility, and much creaking. Bilateral talipes equino-varus, and lordosis on standing only. Knee-jerks present, superficial reflexes normal, sensation, normal. Electrical reactions only quantitatively diminished—no reaction of degeneration. Excellent mentality.

ROCHER, H. L., 1913.—Female, 2½ years old. Extended left lower extremity. Right, adducted, hip flexed 40°, knee flexed 90°. Double pes equino-varus. Arms applied to sides of body and in marked pronation. Acromions point forward. Area of skin over olecranon like a vaccination mark, not adherent to bone. Scapulo-humeral abduction to 30°, few degrees of external rotation, also internal rotation. Forward abduction humerus to 45°. Glenoid surfaces irregular. Elbows fixed at about 90°. Crackling in elbow joints with some lateral mobility. Right radius subluxated anteriorly. Club-hands. A " petite fossette " of skin over the ulna styloid.

CASE 2.—Male, age 6 months. Club-hands. Right elbow extended to 40°. Left, normal. Skin of hands, forearms and about both elbows like " molasses." Lower extremities externally rotated, double congenital dis-

location of the hip. Rigidity of knees, left flexed 60°, right 45°. No lateral movement. Extension normal. Patellæ atrophied, with overlying cutaneous fossette. The skin sensation of the lower extremities similar to upper, but more gelatiniform. Talked of lymphangiectasis of the subcutaneous tissue. Club-feet. Large right inguino-scrotal hernia.

CASE 3.—All joints of superior extremities fixed. Limbs in extension and forced internal rotation. Shoulders adducted. Scapulæ fixed to shoulders. Pronation and supination very limited. Elbows point to front. Crackling in joints. Club-hands with fossette at wrists. Club-feet. Knees extended but flexed to 45°. Slight left genu valgum. Appearance of conic cylinders. No muscle reaction of degeneration.

CASE 4.—Male, age 20 months. Flexed upper extremities, right to 40°, left to 18°. Marked internal rotation. Pronation and supination much limited. Club-hands with some movement. Thumbs opposed. Metacarpo-phalangeal rigidity, extension of fingers very limited. No muscle reaction of degeneration.

GRISEL, 1913 (cit. Rocher).—Age 2½ years. Lower extremities abducted, externally rotated. Knees flexed. Club-feet. Right club-hand, flexed fingers, restricted abduction of thumb. Elbows flexed. Left hand in fixed hyper-extension with extended fingers.

JACOBI, 1877 (cit. Rocher).—Male, age 6 months. Elbows extended and club-hands. Knees flexed, feet clubbed. All voluntary joint movements absent.

KÖRTE, 1876 (cit. Rocher).—Male. Contracted hips and knees, absent patellæ.

DUVAL (cit. Rocher).—Female, age 35 years. Flexed knees, flexed hips since birth. Also flexed elbows.

STERN, W. G., 1923.—Four cases. Completely extended arms and legs. Limitation of all joints of body except maxillary joints and vertebral column. Arms internally rotated, thighs outward. Fusiform or cylindrical shape to elbows and knees. Wrists flexed, hands compressed, feet twisted. Head and neck not deformed. Marked atrophy of muscles of shoulder girdle. One case had a congenitally dislocated hip. Another had subluxation both tibiæ. X-ray studies of bones, negative. Capsules of joints and surrounding tissues unduly thickened. Movement of joints did not set up tension or contractures in tendons controlling joint motion. Musculature of limbs not well developed. In no case was any general loss of nerve or muscle function demonstrable. All reflexes and sensations normal as far as could be demonstrated. One patient somewhat mentally backward. Dimpling of skin over elbows and patellæ.

CAMPBELL, W. C., 1923.—Two cases. Similar clinical manifestations to cases of Stern. One case had anterior dislocation head of radius. Radius 1 inch longer than ulna. Both forearms extremely pronated.

MCCHESNEY, G. J., 1923.—One case. Contractures of elbows and legs. Marked compression deformity of thorax, sternum depressed. A newborn baby.

GAENSLEN, F. J., 1923.—Case similar to Stern's but knees in flexion and movement hips and shoulders very restricted. Three months old. Marked limitation of movement of spine.

LEROY LOWMAN, C., 1923.—Both feet turned completely backward. Club-hands. Complete absence of deltoid muscles. No shoulder abduction.

LEWIN, P., 1925.—Male, 6 weeks old. Normal delivery, born at term.

Six normal siblings. Malnourished. Hands flexed at right angles to wrist with ulnar deviation. Both thumbs in adduction and flexion. Fingers like claws, long and tapering, lying across each other. Arms in extension. Flexion at elbows impossible. Abduction not above 60°. Legs abducted, externally rotated. Much limitation of hips and knees, impossible to extend knees beyond a right angle. Feet in marked equino-varus. Head round, symmetrical. Sutures closed. Anterior fontanelle admitted only tip of little finger. Neck not rigid. Pug-nose. Head circumference 35 cm.

Case 2.—Fourteen months old. Club-hands and club-feet. Extreme talipes equino-varus, bilateral. Legs formed a diamond shape. (No other details.)

Rocher, H. L., 1925.—In a paper " Main Bote Congenitale et Main Bote Paralytique " mentions 3 more cases he had observed since his 4 published in 1913. All had club-hands and club-feet, genu valgum and congenital dislocation of the hip.

Scarlini, G., 1926.—Female, age 7 years. Parents alive and well. No deformity in family. Two siblings. Oligohydramnios in the 3 pregnancies. Patient, the third child. Fœtal movements ceased at 6 months. Born at term, normal vertex presentation. Upper extremities in extension. Shoulder movements limited in all directions, abduction of right, 90°, left, about 100°. Right elbow, flexion to 100°, left to 90°. Supination absent bilaterally. Hands and fingers, relatively normal. Lower extremities in vicious flexion posture, making upright position impossible. Hips, flexed, abducted about 15° and externally rotated about 30°, adduction and internal rotation impossible. Right knee flexed to 65°, left to a right angle. Legs crossed posteriorly with feet in severe equino-varus and pes cavus. Patellæ present radiographically, not clinically. Adventitious bursæ overlying knees, result of ambulation. Very poor musculature of legs. Electrical reactions present, no reaction of degeneration. Roentgen study of bones, normal.

Case 2.—Male, age 22 months. Parents healthy, no familial deformity. Abnormal presentation and difficult delivery. Hips flexed and abducted 45°, externally rotated 90°. Knees, flexed to right angle. Feet in very severe equino-varus position. No active movements of hips, some passive movements in all directions. Retroversion of femoral necks. Clinically absent patellæ. Very severe atrophy of muscles of thighs and legs. Weak electrical responses but no reaction of degeneration. Testes, ectopic in inguinal canals.

Case 3.—Male, age 7 years. Parents well, no familial deformity. Six siblings, one dead of meningitis, others normal. Birth normal despite deformities of feet and knees observed at birth. Lower extremities only affected. Knees in flexion and feet in equino-cavo-varus. Right knee flexed 45°, left 90°; subluxation tibial condyles on femoral—no joint deformity. Full active flexion. Limits of extension as above but force extended joint, passively, a little further on the right but had no effect on the left. Patellæ normal. Very marked atrophy of thigh muscles with reduction in electrical responses but no reaction of degeneration. Large adventitious subcutaneous bursæ of the knees. Feet in very severe equino-varus.

Poli, A., 1927.—Male, age 6½ years. No familial deformities. Father, chronic alcoholic. Five siblings, one died of croup, others normal. Mother suffered from persistent vomiting of pregnancy. Patient born at term, but mother's abdomen during pregnancy said to have been of smaller girth than

it was during other pregnancies. Did not notice fœtal movements until 2 months before term. Upper extremities, poor musculature, slight exaggeration of pronation of forearms. Some limitation of abduction and forward flexion of shoulders, almost normal rotation. Elbow, limit of flexion, 90° on right, 70° on left. Normal extension of right, some limitation of left. Supination diminished on both sides. Flexion deformity of middle and index fingers of right hand. On left, index finger flexed into palm. Flexion due to contracture of soft parts. Lower extremities, both externally rotated 90°. Popliteal fossæ face each other. Abduction of right, adduction of left, hip-joints. Marked hypoplasia of muscles. Right lower extremity appeared a little shorter than left. Flexion of hips normal. Right knee, flexion 45°; left, no flexion. Right, extension normal; left, hyperextended 10°. Lateral movement possible. Subcutaneous and muscle tissue poor. Electrical reactions present, no reaction of degeneration. Feet in marked equino-cavo-varus. Left genu recurvatum and valgus position. Scoliosis with tilt of pelvis. Dislocation of right hip, absent left patella, hypoplasia of right. Left coxa valga. Stresses periarticular structures as being primarily responsible for joint deformities.

Rocher, H. L., and Ouary, G., 1929.—Female. Very short, thin upper extremities. Arms internally rotated, olecranons pointing forward. Forearms fully extended and pronated so that palms open to the front. Hands in cubito-palmar club-hand position, with flexion of metacarpophalangeal and interphalangeal joints. Hunched shoulders ("poupeé en bois" of Redard). Muscle groups very atrophic, even non-existent, but triceps muscle and flexors of fingers, normal. Scapulæ moved with shoulders, only 45°. Shoulder-movement backwards and forwards also only 45°. Elbows flexible 30° with crackling in joint. Lateral mobility about 30°. Forearm much pronated, and attempted supination painful. Also crackling at wrist with movement. Carpal bone prominent. Marked bilateral club-foot. X-rays showed some possible atrophy of humeral diaphysis.

Case 2.—Age, 3½ months. Lower extremities in extension. Feet in varus position. Short lower extremities with umbilicated fossæ over the trochanters, and conical in shape with excess subcutaneous tissue, making palpation of muscle-groups difficult. Hips, semiflexed on pelvis, but thighs not able to touch pelvis. Knees flexible 20° but habitually in extension. Very small kneecap. Lumbar gibbus. X-rays showed imperfect formation of lumbar vertebræ. Bilateral congenital dislocation of hip and total aplasia of sacrum. Undeveloped centres of ossification of talus and calcaneus.

Berntsen, A., 1930.—Female, age 29 years. Congenital ulno-palmar deviated club-hands with contracted fingers, and thumbs in adduction and opposition. Flexion contracture of both elbows.

Case 2.—Female, 19½ years. Family history negative. Similar to first case as far as hands, fingers and thumbs concerned, but elbows contracted in extension to 180° making eating extremely awkward, in that flexion in right possible only to 90° and left to 170°. Shoulder movements limited to ¾ normal range. Atrophy of both upper extremities. Bones and skull normal by Roentgen examination.

Case 3.—Male, age 20½ years. No deformities in family. Contractures of hand, pes varus, genu valgum, scoliosis.

Rocher, H. L., and Laporté, A. 1931.—Infant, age 1 month. Breech delivery.

Superior Extremities.—Posterior subluxation of shoulders. Left elbow rigid—flexion limited to 90°. Cubito-palmar club-hands without bony deformity. Muscles of arms (biceps, triceps) and posterior muscles of forearm, almost absent.

Inferior Extremities.—In extension. Knees not flexible. Right lower extremity in marked external rotation. Severe pes equino-varus. Marked atrophy of muscles especially quadriceps, femoris and antero-lateral aspect of legs.

Sheldon, W., 1932.—Elbows in rigid extension, forearms fully pronated, wrists flexed. All movements of shoulders performed but abduction slow and limited, scapulæ move with humeri. Passive movement elbows only 15°. Forearm fully pronated could be semi-supinated. Wrists had only ¾ normal range of flexion, could not be cocked. Thumbs and fingers, normal. Arms tapered, circumference least at shoulders. Rigid extension of knees and bilateral pes equino-varus. Full passive movement of hips. Right knee to right-angle, left to ¾ right angle. Active movement a little less. Crackling. Movement stopped suddenly at end of range, *cf.* fibrous ankylosis. Limited ankle movement with short Achilles' tendons. Toes fully extended passively. Legs also tapered. No spasm detected under anæsthesia. No sensory or trophic changes, or muscle reaction of degeneration.

Krause, A., 1932 (cit. Kallio, K. E., 1948).—Contributed 3 cases. One case, contractures in upper limbs only, and a wedge-shaped cervical vertebra. Other cases had also contracture deformities lower limbs with congenital dislocation of hip and pes equino-varus.

Price, D. S., 1933.—Little or no amniotic fluid at birth, which was otherwise normal. No abnormalities in family. No voluntary movement of shoulders or elbows. Elbows thickened and spindle shaped. Left elbow flexed to a right angle. Only slight flexion in right. Congenital club-hands with ulnar deviation. Wrists flexed and not fully extensible. Finger joints flexed and partly overlapping and slightly webbed, could be separated and extended with difficulty. Skin on palmar surface tight and contracted. Legs everted and stiff. Coxa vara and thighs adopt scissor position. Some thickening of knees but they could be flexed and extended passively. Spine straight and rigid, due to spasm which disappeared later. Congenital dislocation of hips. No other bony deformity by Roentgen studies.

Middleton, C. S., 1934.—Female, age 7 years. Healthy, unrelated parents. Several healthy siblings. No family history of deformities. Bilateral club-hand, fingers slightly flexed, thumb adducted, digits thin with wasting of interdigital tissues giving a false impression of webbing. Powerful flexion of fingers, extension wrist and fingers weak. Forearms in full extension, no active flexion. Passive, to 70°. Paralysis of triceps, brachialis and biceps muscles. Region of shoulders relatively normal but active abduction, weak. All muscles of limbs wasted. Roentgen study, normal. Marked equino-varus of feet with wasting. Knees flexed to right angle. Hamstrings weak, quadriceps muscle had some power. Pelvic girdle muscles, weak. X-rays showed normal hips. Bright and intelligent child.

Case 2.—Female, age 8 years. Three normal siblings. Parents healthy and unrelated without family history of deformity. Bilateral ulnopalmar club-hand. Slight active flexion of fingers. Elbows fixed rigidly in extension from birth. Much wasting of arm and shoulder muscles. Shoulders fixed in adduction and medial rotation. No active or passive abduction. Wasting

of superior scapular muscles. Difficulty in using accessory muscles of respiration, when crying in infancy, this gradually passed off.

Case 3.—Female, age 6 months. Slight spastic contracture of wrist at birth, easily overcome and disappeared completely in a few days. Feet in extreme talipes equino-varus. Knees in hyperextension, only a trace of passive flexion. Hip joints flexed. Muscle wasting marked. Plenteous layer of subcutaneous fat. Both hip-joints dislocated. Slight microcephaly, defective mentally.

Case 4.—Male, age 8 weeks. Healthy unrelated parents. Four normal siblings. No family history of deformities. Well-marked equino-varus. Lower limbs short and wasted. Knees extended, passive flexion to some 10° only. Some power within this range. Movement of hips slightly limited in all directions. Bilateral dislocation of hips, irreducible.

Case 5.—Male, age 3 weeks. First born. Healthy parents. Easy labour. Arms fully extended at sides. Slight movement only. Pectoral muscles acted fairly well. Passive abduction slight, not active. All scapular and deltoid muscle wasted. Pointed shoulders. Elbows moved passively to right angle, no active flexion. After passive flexion, triceps muscle able actively to extend elbow. Ulno-palmar club-hands, wasted and weak wrists. No bony defects. Extension of fingers poor but good grip possible. Thumbs adducted. Lower extremities straight and movements hip-joints very limited. Passive flexion of extended knees, to right angle. Hamstrings did not act but some power in quadriceps muscle. Active extension from flexed position good. Bilateral extreme talipes equino-varus. Left hip congenitally dislocated, right, exaggerated coxa vara.

Case 6.—Male, age 7 years. Normal I.Q. Family history negative. Dry labour. Both arms thin and weak. Shoulders normal. Elbow movement limited to 90° on left and 10° on right. Both forearms pronated, pronation and supination very restricted, almost absent on left. Bilateral ulno-palmar club-hand. Knees flexed and feet in talipes equino-varus. Right hip dislocated. Extreme dextro-convex dorsilumbar scoliosis but no bony abnormality or aplasia.

Moncrieff, A., and Wiles, P. 1934.—Male, normal pregnancy. Liquor amnii normal. Scapulæ elevated producing a very short neck. Restricted shoulder movements. Movement right elbow, full, left elbow could not extend beyond right angle. Wrists normal. All fingers flexed, could not extend. Deformity of metatarso-phalangeal joint of thumb. Hips extended to 40°. Right knee extended to 45°, left knee to right angle. Extreme valgus feet. Roentgen studies showed scapulæ and thumb bones in abnormal position and high position of left greater trochanter. Upper part of chest somewhat compressed. No reaction of degeneration.

Schapira, C., 1935.—Seventeen cases, one of which had already been reported. Ten males, 7 females, aged 8 days, 18 days, 20 days, 22 days, 25 days, 1 month, 45 days, 2 months, 3 months, 3 months, 4 months, 5 months, 6 months, 6 months, 8 months, 10 years, 13 years. No evidence of familial or hereditary traits. Pregnancy normal in 15. One mother had repeated hæmorrhages during pregnancy, the other had weak fœtal movements. Three cases came of mothers who had borne many normal children. Three cases had been preceded by abortions. Three were associated with oligohydramnios, one with polyhydramnios, dystocia occurred 8 times, podalic presentation occurred 5 times. Three cases of Rocher's type I (4 limbs in extension),

1 case upper limbs extended, lower limbs flexed. Six cases had only lower limbs affected and remaining 7 had various combinations of extension and flexion of different joints and adduction and internal rotation of arms; extension of elbows, pronation of forearm. Only 2 cases with elbows in flexion with limited extension. Hands were involved in 10 of 17, typical congenital club-hands. Palmar flexion and cubital deviation. In only 1 case hand fixed in dorsal hyperextension. In all cases with deformity of hands and fingers the thumb was fixed in adduction and opposition with limitation of abduction. In lower extremities with rigid hips there was always external rotation, with knees in rigid extension in 13 cases (sometimes hyperextended with valgus deviation). Only 3 cases with knees fixed in flexion. Patellæ not detectable in 9 cases, present but difficult to detect clinically in some cases. Sixteen cases had pedal deformities, 11 with typical bilateral pes equino-varus, 3 with pes valgus and 2 with pes equino-varus of one foot and pes valgus of the other. The integrity of the skeleton was noteworthy. There was articular abnormality of knee once, recurvature of lower segment of tibia once; twice, deformity of bones of feet. Subluxation of hip joints in 7 cases. The tendons, ligaments and articular capsules constantly involved.

Gerri, A., 1935.—In an article entitled congenital multiple deformities this author gives details of 4 cases. The first three can unhesitatingly be classed with the syndrome under consideration.

Case 1.—Female, 5 years old. Father deceased, was alcoholic, mother well. Right hand in forced adduction. Thighs externally rotated 90°, valgus knees, rigid and flexed 20°. Feet adducted 45°, supinated about 75° and in 90° equinus. Active abduction of hips and external rotation limited. Normal flexion and internal rotation. All passive movements possible. Very slight limitation of movement of knee. Bilateral congenital dislocation of hip.

Case 2.—Female, 9 years old. Family history negative. Bilateral club-hand, bilateral valgus hips, bilateral pes equino-varus. Knees acutely flexed and subluxated. Active and passive extension impossible.

Case 3.—Male, 4 months old. Family history negative. Difficult birth, instrumental. Bilateral deformed hands, bilateral dislocation of hip, rigid and valgus knees. Bilateral club-feet. Umbilical hernia and inguino-scrotal hernia, sacral spina bifida.

Cook, L., 1936.—Female, 7½ years. Mother had " some hæmorrhage after a long omnibus ride " in the third month of pregnancy. Vomited much in the sixth month. Rapid delivery at term, an unreduced R.O.P. with extended legs. Normal amniotic fluid. Legs appeared " back to front " at birth, genu recurvatum and severe equino-varus and cervical opisthotonos. Poor musculature. Had been orthopædically treated by this time, when appeared typical mongolian idiot, with abnormal mobility of all joints except knees and ankles. No detectable heart lesion but peripheral circulation, poor. Frequent chilblains. Patellæ not palpable. X-rays showed subluxation lower end of femur. Patellæ faintly seen.

Horeyseck, L., 1938.—Male, age 4 months. Florid rickets. Right arm hung vertically. No active movement. Full passive movement of right shoulder. Left shoulder ankylosed. Right elbow fixed in extension with 15° passive movement. Left elbow also extended, no active movement, but passive to 90°. Both hands clubbed in marked ulnar deviation. Fingers flexed, thumb folded into palm. Could not grip or move fingers of right

hand. Hips and knees normal. Illustration shows upper extremities like stuffed sausage skins.

CASE 2.—Male, 2 years old. Evidence of rickets. Could neither sit nor stand nor walk. Both arms extended at elbows. Stiff left shoulder joint, limited active movement. Passive, to over 90°. Right shoulder movement good, active to over 90°. No active movements of elbows. Hands in 110° volar flexion, and ulnar deviation. Fingers slightly flexed and thumb opposed. Fingers of right hand could be used but poor grip. Congenital dislocation left hip. Right genu valgum. High grade clubbing of feet. Patellæ not palpable. Roentgen study normal except for the dislocation.

ALTMAN, H. S., and DAVIDSON, L. T., 1939.—Age 2 weeks. Mother had normal pregnancy and confinement. Rather spastic and unable to raise upper eyelids. Increase of spasticity. At 8 months inability to extend legs, flex fingers. Marked ptosis of eyelids. Spastic. Skin like scleroderma. Fingers rigid. Legs could not be extended passively. Appeared mentally retarded but this not confirmed later. Bilateral club-foot. Contractures of hamstrings. Distal parts of extremities, cold. Pneumoencephalograms did not warrant diagnosis brain atrophy. Cerebrospinal fluid, normal.

MORENO, M. R., and GESER, R. F., 1939.—Female, age 13 years. Family history essentially negative. Podalic presentation, obstetrical fracture of left femur. *Superior extremities.*—Some limitation of abduction of wrists. Atrophy of dorsal interosseous muscles. Second and fifth fingers in fixed flexion, and thumbs in fixed opposition and flexion at the metacarpo-phalangeal joint. Extension and flexion of index finger, difficult. *Inferior extremities*: Cylindroid in appearance and in fixed extension. Skin suggested peripheral vasomotor disturbance. Abundant subcutaneous tissue in some regions obscuring the outline of the subjacent muscles. Thighs externally rotated, bilateral congenital dislocation of hip-joints. Knees extended. Superficial skin depressions at outer borders of patellæ. Longitudinal sulcus running parallel to tibial crests, not compatible with normal anatomy of the region. Feet, externally rotated with prominent talus. Different degrees of active and passive movement in the hips, knees, ankles and feet. Patient's standing equilibrium unstable and associated with a lumbar lordosis. X-rays showed bony abnormalities of minor degree in pelvis, knees, and feet and subluxated patellæ. No reaction of degeneration. Serology negative for mother and child.

CASE 2.—Male, 11 years old. Slight flexion of fingers, no full active extension. Thumbs fixed in opposition and flexion at the metacarpo-phalangeal joint. Tendency to hyperextension at interphalangeal joints and limitation of flexion. Lower extremities showed slight degree of fixed flexion of knees in erect position. Incurving of both tibiæ. Patellæ normal. Bilateral pes valgus. Subcutaneous tissue abundant in thighs but minimal in legs. On internal border of the patellæ were depressions resembling scarring from healed osteomyelitis fistulæ and over the anterior aspect of tibiæ were oblique sulci running down to the foot and across the tibial muscles. X-rays showed slight degree of coxa vara. No reaction of degeneration of muscle.

BADGLEY, C. E., 1943.—In his Presidential Address mentioned 5 of his cases. His cases included internally rotated upper and externally rotated lower, limbs. Absence of normal creases separating trunk from limbs. Muscles atrophic and degenerated. Congenital dislocation of hip-joint present with malformation of sacrum.

KATZEFF, M., 1943.—Collected 18 case reports in Children's Hospital,

Boston, Mass. Ten males, 8 females. Ages from 5 days to 10 years 11 months. No history of deformity in relatives or parents. In one case maternal uncle, age 20 years, had never walked. Regions involved: hands and feet; hands, feet, knees and hips; hands, elbows, feet and knees; feet, knees and dislocated hip; hands, feet, elbows, knees and hips; hands, feet, hips both dislocated; feet and dislocated right hip; feet, knees, hips and spine; feet, knees, hands and dislocated hips; feet, hands, elbows and dislocated hips; feet, knees, hands, hips and spine; feet, hands, knees and shoulders; feet, hips and spine. Five cases had dislocation of the hip, bilateral in 3. Two had cryptorchidism. Three had inguinal hernias. One had a cleft palate. The extremities lacked the usual contours.

EALING, M. I., 1944.—Case complicated later weeks of pregnancy and delivery. Uterine inertia from hydramnios. Spastic deformity of all limbs with movement limited in all directions, at shoulders, elbows and wrists. Elbows semi-flexed, also wrists. Hands with long claw-like fingers being pronated. Thighs and knees in semi-flexion. Severe bilateral talipes equino-varus. Rigidity of temporo-mandibular joint making it impossible to close mouth. No bony lesion. Skin dry and scaly resembling ichthyosis.

DALMAIN, W. A., 1947.—Severe contractures of all the extremities. Both hip-joints dislocated.

CASE 2.—Male, age 9 years. Club-feet. Contracture of both elbow joints and both hands. Coxa vara of hips and flexion deformities of knees.

CASE 3.—Male, sixth of seven children. Difficult delivery. Upper limbs internally rotated and elbows extended. Contractures of both axillæ. Lower extremities normal, except for mild varus deformity of one foot.

BRANDT, S., 1947.—Male, 3 weeks old. Amniotic fluid, normal. Sibling normal. Unable to suck, loss of weight. Skin dry and flabby, scaly. Subcutaneous tissue totally absent. Arms inwardly rotated, with backs of hands turned forwards. Hands in 45° volar flexion, slight ulnar deviation. Hands flexed passively still further but no passive extension. Thumb opposed and fixed. Elbows flexed passively to 70° from full extension. Only a few degrees of pronation and supination. Abduction of shoulder joints, with scapulæ, to 45° only. Rotation very slight. Deltoid and latissimi muscles, short and tight. Condition on the whole symmetrical in the upper extremities. Muscle atrophy of arms, pronounced; biceps, a string, pencil-thick. Forearms thickened with a doughy, œdematous subcutaneous tissue. Abduction left hip-joint to 30°. Left knee, passive flexion to about 40°. Left hip to 30° and rotation about 10°. Left foot turned inwards at ankle joint. Plantar surface of heel faced medially, dorsum of foot, anteriorly. Right hip resembled left. Right knee in genu recurvatum. Flexion not possible. Patellæ present. Muscle atrophy in extremities equally distributed. Active mobility of buttocks, practically nil of thighs and legs. Tendon reflexes could not be evaluated because of joint stiffness. Plantar reflexes, normal. No dorsal trunk muscles could be felt. Severe atrophy of cervical muscles. Neck in opisthotonos. Also facial muscle atrophy, especially orbicularis oris and risorius. Mouth closed with difficulty. Lips retracted, gums uncovered. Canine region, lower jaw, prominent. Lower jaw, slightly wry, left side higher than right. Hypospadias. No reaction of degeneration. Cerebro-spinal fluid, normal cell count and protein. Excretion of creatinine, 2·5 mg. per day.

ROSSI, E., 1947.—Reports 8 cases. In 2, all 4 extremities affected. In 4, lower limbs only affected. In 2, upper limbs only affected. In one,

mandibular articulation affected and in another the spine was involved. Muscle hypo- and aplasia in all cases. Subcutaneous changes present in all. Hypoplasia of patellæ in two. Thymus hyperplasia in one. Hypoplasia of the mandible in four. Roentgen studies showed a generalised osteoporosis but normal shape of the bones. One case had unilateral ptosis of the eyelid. One had synechiæ of the lobes of the ears. Two had cleft palate. One had a left facial palsy. Interdigital webbing was a constant finding.

Kallio, K. E., 1948.—Female, age 39 years. Nine normal siblings. No familial abnormalities. Club-feet and rigid hands. Upper limbs relatively short and inwardly rotated. Atrophy and limitation of movements of shoulders. Elbow range 100°-135°. Rotatory action impossible. Left forearm in pronation, right in mid-position. Left radius 0·5 cm. longer than right with subluxation of capitulum. Hands clubbed, fingers cramped, thumbs adducted. Marked lumbar lordosis. Congenital bilateral dislocation of the hips. Thighs rotated externally. Knee movements 170°-160°. Ankle movement restricted. Feet short and broad. Sensation and reflexes, normal. Fossæ semilunaris ulnæ considerably deeper than normal. Hypoplasia of heads of humeri and of the handbones. Left radius gave impression of being too long. Asymmetry of 11th and 12th dorsal vertebræ.

Case 2.—Female, aged 11 years. Marked atrophy of shoulder girdle and upper limbs. Arms raised actively 40° anteriorly, not at all laterally. Passive raising to 70°. Rotation 20°. Elbows fully extended but passive flexion to 90°. Forearms in mid-position. No pronation, slight supination. Severe club-hands with 10° active flexion possible. Fingers semi-flexed, no passive flexion. Thumbs adducted. Slight limitation of flexion of hips. Knees, normal. Severe club-feet. Roentgen examination, normal.

Case 3.—Female, 3 years old. Shoulders atrophied. Arms inwardly rotated. Passive movements of shoulders and elbows restricted. Club-hands, wrists in ulnar-volar flexion. Congenital dislocation of left hip. Thighs outwardly rotated. Slight limitation of extension of right knee. Club-feet. Roentgen examination, negative.

Case 4.—Female, 2 years old. No abnormal family history. Breech delivery. Marked atrophy of shoulders and upper limbs. Range of elbow movement, 135°-180°. Club-hands, wrists in volar flexion. Fingers cramped. Congenital dislocation of right hip. 40° flexion contracture of left hip. Right knee in 45° valgus position and left in corresponding varus. Right knee movement, 30°. Motion in left knee, good. Short rigid club-feet. Roentgen study, negative.

Bearán, M. S., 1948.—Male, 8 years old. Hutchinson's teeth. Deltoid muscles totally atrophied. Pectorals contracted. Rigidity and loss of abduction of arms but anterior elevation normal. Posterior elevation diminished. Elbows almost completely rigid. Cutaneous dimples over posterior aspect. Club-hands with flexion middle finger. Hips slightly rigid. Left knee rigid in semi-flexion. Right knee rigid in full extension. Both legs internally rotated. Bilateral club-feet.

Case 2.—Male, 6 years old. Poor intelligence. Frölich type, genital atrophy and excessive deposit subcutaneous fat. Large hernia on right due to abdominal muscle aplasia. All extremities rigid. Deltoids atrophied, contraction both pectorals, elbows in extension, hands in flexion and thumbs adducted. External rotation both femurs. Full flexion both knees and bilateral club-feet.

Case 3.—Male, aged 6 months. *A twin, the other normal.* Atrophy of deltoids marked. Contraction of pectorals. Elbows extended with very slight flexion. Club-hands with cubital deviation. Right scapulo-humeral movement much restricted. Cutaneous fossettes over trochanters of femurs which were markedly externally rotated and rigid. Lower limbs had conic-cylindroid appearance with little flexion of knees and bilateral pes equino-varus.

Case 4.—Male, aged 4 years. Mother had normal pregnancy and delivery. There had been little intra-uterine movement at seventh month. Pale, elastic skin a little adherent to deeper tissues. Atrophy and paralysis of deltoids and biceps. Bilateral contraction of pectorals. Elbows in extension with some flexion in right only. Forearms pronated. Club-hands. Thumbs adducted. Left hip free but left knee rigid at angle 155°. Foot in equinus. Right hip free, knee rigid in extension, foot in equinus.

Case 5.—Male, aged 8 years. Elbows rigid with some flexion. Each thumb with 3 phalanges, with right distal joint acutely flexed. Short index finger. Hips normal, knees rigid in marked flexion—legs atrophied and cylindroid. Both feet with polydactyly—6 toes on each foot.

Case 6.—Female, aged 7 years, much adipose tissue. Cutaneous fossettes. Asymmetry of face. Strabismus left eye. Short neck, resembling Klippel-Feil. Atrophy of deltoids, contraction of pectorals. Excursion of elbows 30°. Thumbs contracted into palm. Middle finger in semi-flexion, without extension, but some active flexion. External rotation marked in lower extremities. Bilateral dislocation of hips. Rudimentary patellæ. Knees in rigid extension without active or passive flexion. Rigid pes valgus bilateral.

Case 7.—Female, 19 years old. Mother with lues. Asymmetry of face with wry lower jaw, teeth characteristic of heredolues. Neck like Klippel-Feil. Kypho-scoliosis. Atrophied deltoids. Club-hands, fingers is in Volkmann's contracture. Hip movement much limited—semi-rigid in intermediate flexion. Knees rigid—semiflexed. Pes equinus. This patient had previously been operated on.

Arce, G., and Pereda, F. M., 1949.—Female, 3 days old. Mother had had abortion previously, at 3 months. Pregnancy normal, rather long labour. Small, receding mandible. Pterygium-like skin folds extended from chin to thorax. Slight hypertelorism. Elbows semiflexed and impossible to extend completely. Forearms pronated and difficult to supinate. Wrists flexed in radio-varus, difficult to extend. Thumb and index-finger extended, other fingers hyperextended at metacarpo-phalangeal joint and flexed at inter-phalangeal joint. Symmetrical disposition. Bilateral metatarso-varus deformity of feet. X-rays showed changes from the normal in the long bones of the upper extremities.

Case 2.—Female, 24 hours old. Cæsarean baby. Serology negative but mother's Kahn reaction positive. Normal pregnancy. Limitations of all movements of upper extremities. Fingers flexed, extension rather difficult. Thighs flexed on pelvis 100-110°. Limitation of all movements at hips. Knees in almost complete extension, could be flexed to 160°. Subcutaneous tissue rather hard giving fusiform aspect to limb and obscuring bony outlines. Ankle-joints stiff. Feet in equino-varus. Syndactyly of second and third toes bilaterally. X-rays showed minor deviations of bones from normal. C.S.F. normal in appearance and pressure. Albumin 0·45 G/per cent., globulin positive.

Case 3.—Female, 8 days old. Negative serology. Umbilical hernia.

Lumbo-sacral meningocele. Thighs and knees not completely extensible. Severe bilateral pes equino-varus. Both hip-joints dislocated and thighs externally rotated. Apparent swelling of knees. X-rays showed minor changes in the bones.

OLEAGA, F., and MUGURUZA, J., 1950.—*Male twin*, age 5 years. Co-twin normal. Parents healthy but their relatives included one with prostatic adenoma, and one who died at 21 years in diabetic coma, and another with acromegalic oligophrenia. Three siblings of the patient, one with unilateral cryptorchidism. Mother had anorexia during first trimester of pregnancy and was involved in an accident in the sixth month. She had intense œdema at the end of pregnancy. Patient born normally and second-born. Placenta single but of great volume, bichorionic and biamniotic. Patient had hypospadias and bilateral cryptorchidism. *Superior extremities*: cylindroid with obscured muscles. Great increase of fat panniculus. Palpation revealed atrophic muscles. Great limitation of flexion of elbows, especially the left, both being held in extension. Wrists in forced flexion with lateral deviation. Positions of hands identical and fingers had normal movement. *Inferior extremities*: cylindroid with absent muscle outlines. Limitation of movement in hips, and much more limitation of knees. Rigidity of tibiotarsal joints. X-rays showed absence of epitrochlear centre of ossification in humerus, and missing left os talus removed surgically.

BASTOS-ANSART, M., 1950.—A general discussion on 30 cases diagnosed from the histories of cases seen in the course of years. Females predominated in ratio of 4 to 1. Attributed to a fibrous tissue degeneration of muscles, and other soft tissues also usually affected. Refers to the possible transitory nature of the affliction.

METCALFE, R. H., 1951.—Female, 1 month old. Arms extended and adducted across front of chest. Forearm fully pronated. Wrists flexed (*cf.* bilateral Erb's palsy). Marked hyperextension of knees and marked talipes calcaneo-valgus of right foot.

In 1905 Rosenkranz collected 55 cases of club-hand of which 35 had associated club-foot, 16 had rigid elbows, and 14 contractures of the knees. It remains uncertain which, if any, of these could be included in the syndrome.

By 1913 Rocher was able to collect 26 cases from the literature and added 4 cases of his own, to make a total of 30. Rocher (1913) quotes Fournier (1912) who reported on 10 cases in his thesis but it is not certain whether these cases were Fournier's own. He also quotes Kirmisson (1912) who observed a surgically treated case.

Stern had seen besides his own cases, several similar to his at the clinic of Dr Chatterton at Phalen Park, St Paul.

Horeyseck mentions Jules Guerin as having described the first case of the syndrome in the middle of the nineteenth century, and Schulte as having collected 33 and adding 2.

Batten (1904) published his case of " myositis fibrosa " which some authors have looked upon as a case of m.c.a.r., but although the final outcome of his case resembled the condition closely in that it had a curved back, a head flexed onto the chest, the face turned to the left because of sterno-cleido-mastoid muscle contracture, flexed arms which could not be freely abducted, short pectoral and biceps muscles, flexed knees with some extension possible, hips flexed in abduction, great toes shorter than the second, third, and fourth toes, and failure of the muscles to relax under anæsthesia, despite these we cannot

accept the case unreservedly and should agree with Batten that " the gradual onset (it began at 9 months of age), the slow progress of the disease, the fixed position which the trunk and the legs gradually assumed, form a clinical picture which bears a striking resemblance to myositis ossificans " and he was inclined to place his case with the inflammatory diseases of muscles.

In 1932 Sheldon found 44 cases in the literature, including one case of Marconi (1922), contributed 4 unpublished cases of Fairbank and one of his own, making 49 in all. Fairbank's cases showed :—

(i) Left wrist fixed in dorsi-flexion and ulnar deviation. Right hand in palmar flexion and ulnar deviation. Limitation of extension of the flexed left elbow. Congenital dislocation of the hip. Left knee extended. Left patella absent. Bilateral club-foot.
(ii) Elbows in extension. Some limitation of external rotation of humeri.
(iii) Elbows and knees in extension. Wrists and hands flexed. Bilateral club-foot. Bilateral congenital dislocation of the hip.
(iv) Bilateral club-foot. Flexed and abducted hips and flexed knees.

E. v. d. Osten-Sacken described 5 cases at the orthopædic clinic of the Academy of Military Medicine in Leningrad, which were very briefly abstracted without details by F. Michelsson (1928).

Weber refers to 5 unpublished cases of Ogilvie, and to a case of Edwards (1938) who demonstrated an infant with " webbing of lower limbs, associated with congenital bilateral contractures of the flexor muscles of the elbow and wrist," and suggests that it is an allied condition. He also refers to the case of Herson (1947) whose case, aged 61 years, had an associated hyperostosis frontalis interna and had had a headache for " as long as she could remember." He alludes to the case of Williams (1947), a boy aged 14 years who had a maldevelopment of the osseous musculature, and subcutaneous tissue, and central nervous system dysfunction.

Rossi tells of Scarzella who showed a case at the Institute of Pathology, Turin, and of Scherer's case at the Hospital of Winterthour. He also tables Lebourg and Lange (1933) as having contributed 2, de Toni (1936) 1, Carnevoli (1938) 10, Fornari-Micott (1938) 2, Heijbroek (1941) 3, and Nicod (1946) 2, cases. Rossi gives no details and the papers have not been available to me. Arce and Pereda (1949) mention Sainton, and Longnet as having written on the subject in 1893 and also quote Marique (1945), Heybroek (1946) and Crisalli (1948) as having published 5 cases between them.