In the name of

GOD

Case Report

Proximal muscle weakness with winging of scapula

M.ILKHANI *MD Professor of neurology*
H.Pakdaman *MD Professor of neurology*
Y. Kholghi *MD*

Department of Neurology,Loghman Hospital,Shahid Beheshti University of Medicine,Tehran,Iran

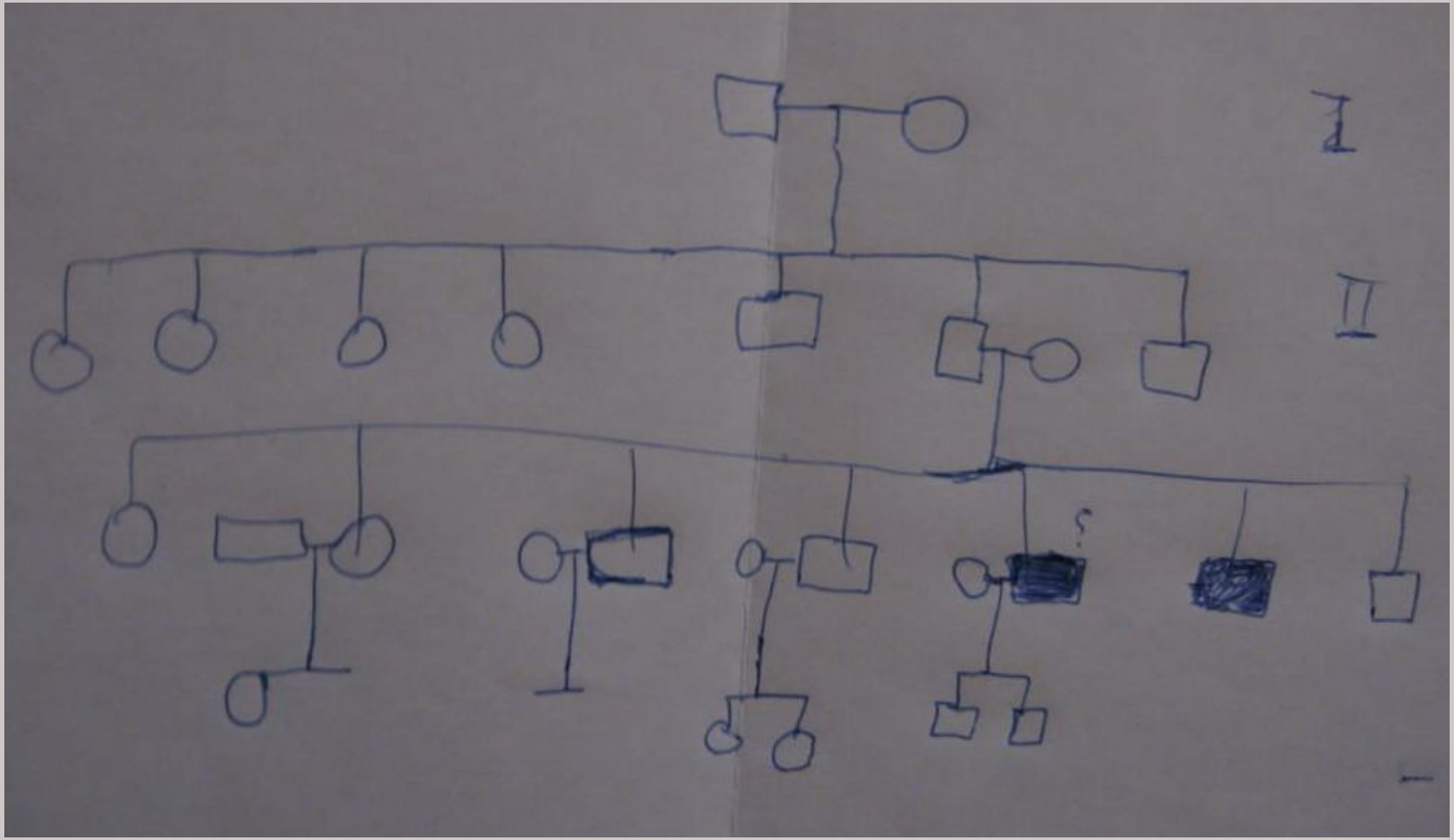
Brief history

- A 20-years old right handed unmarried male.
- He has history of difficulty in displacing of heavy objects and elevation of upper limbs with jutting backward of shoulders, inability to whistling and loss of weight since 5-years PTA.
- He have not complain of weakness in lower limbs and no history of muscle pain.
- He have respiratory complain (dyspenea) at exertion, but no history of difficulty swallowing, and pain and paresthesia in body or fluctuation in weakness.
- No family history similar disease and PMH of medical or surgical diseases.
- History of body building sport IM injection of hormone.

Brief N/E

- Weakness in forceful closing of eyes, inability to whistling with mouth, prominent weakness in shoulder abduction with some atrophy of shoulder girdle (preserved deltoid) and arm muscles, and generalized thin appearance without weakness and atrophy in hip and thigh muscles and no Gower's sign or foot drop.
- STM, forearm, hand, foot, calf, deltoid and MSR was normal and fasciculation and myotonia were not seen .
- Eyes and ears and other N/E was normal.

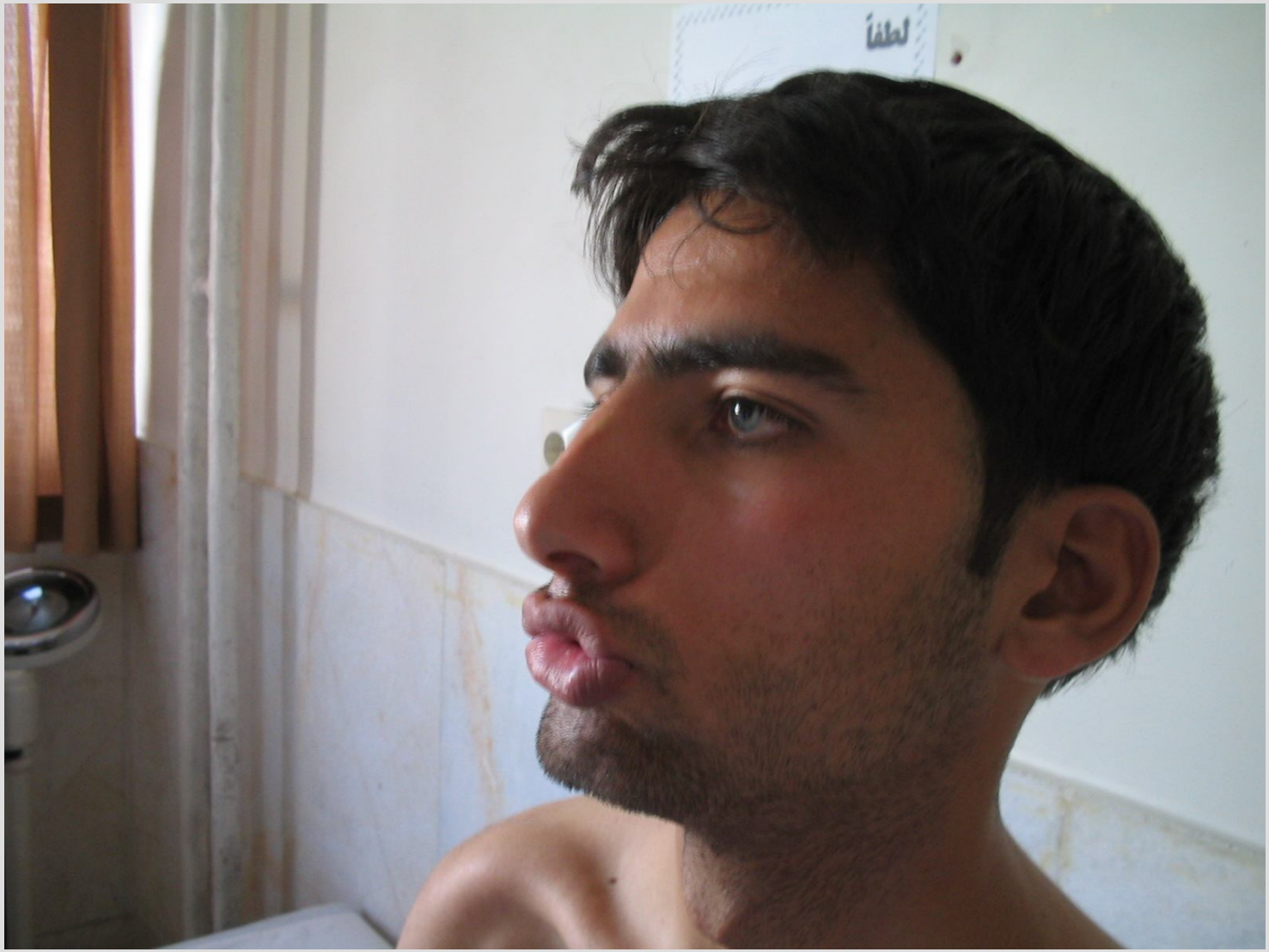
Pedigree demonstrating no family of similar case



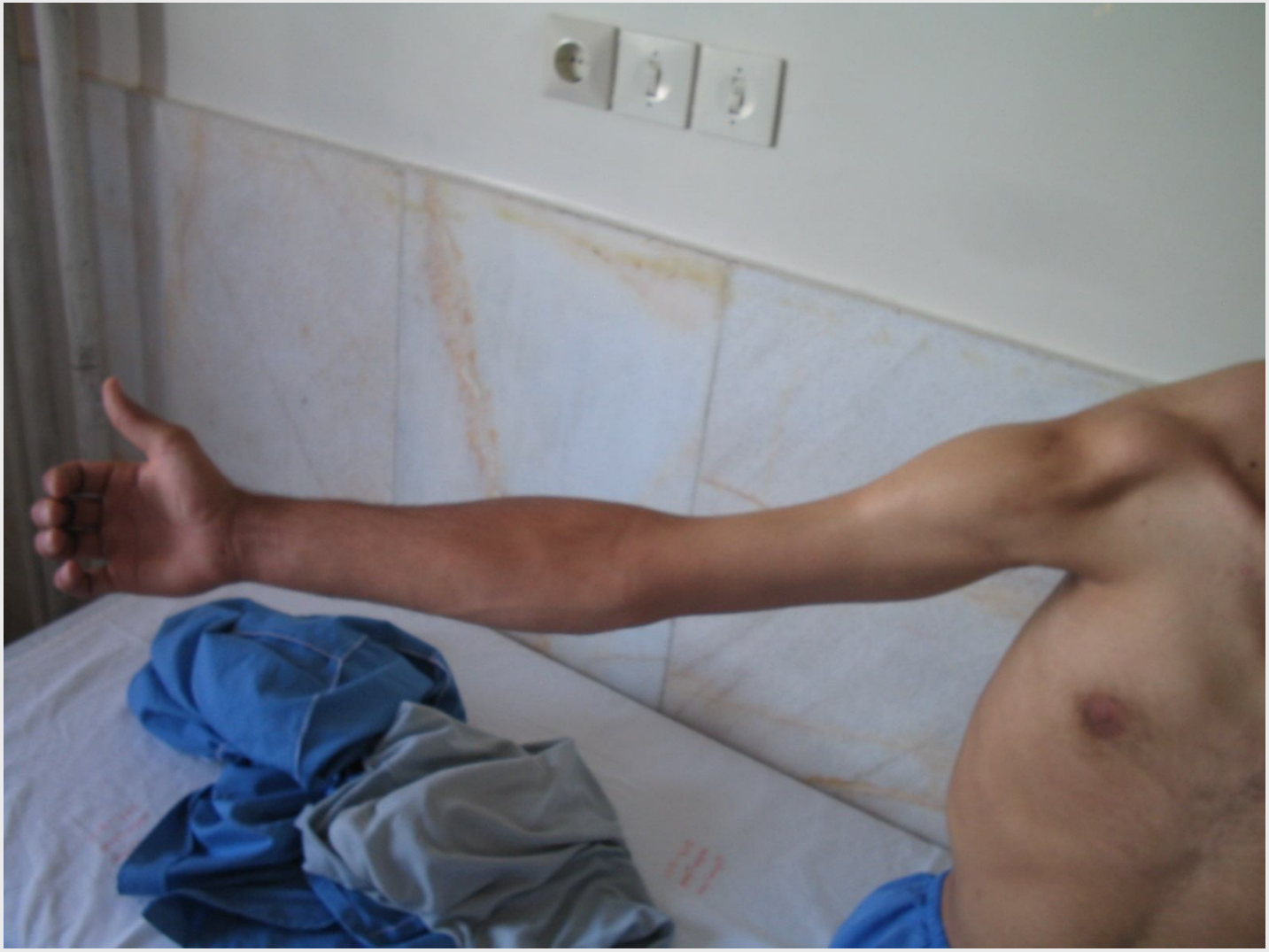
Some special signs in muscle exam

Facial weakness

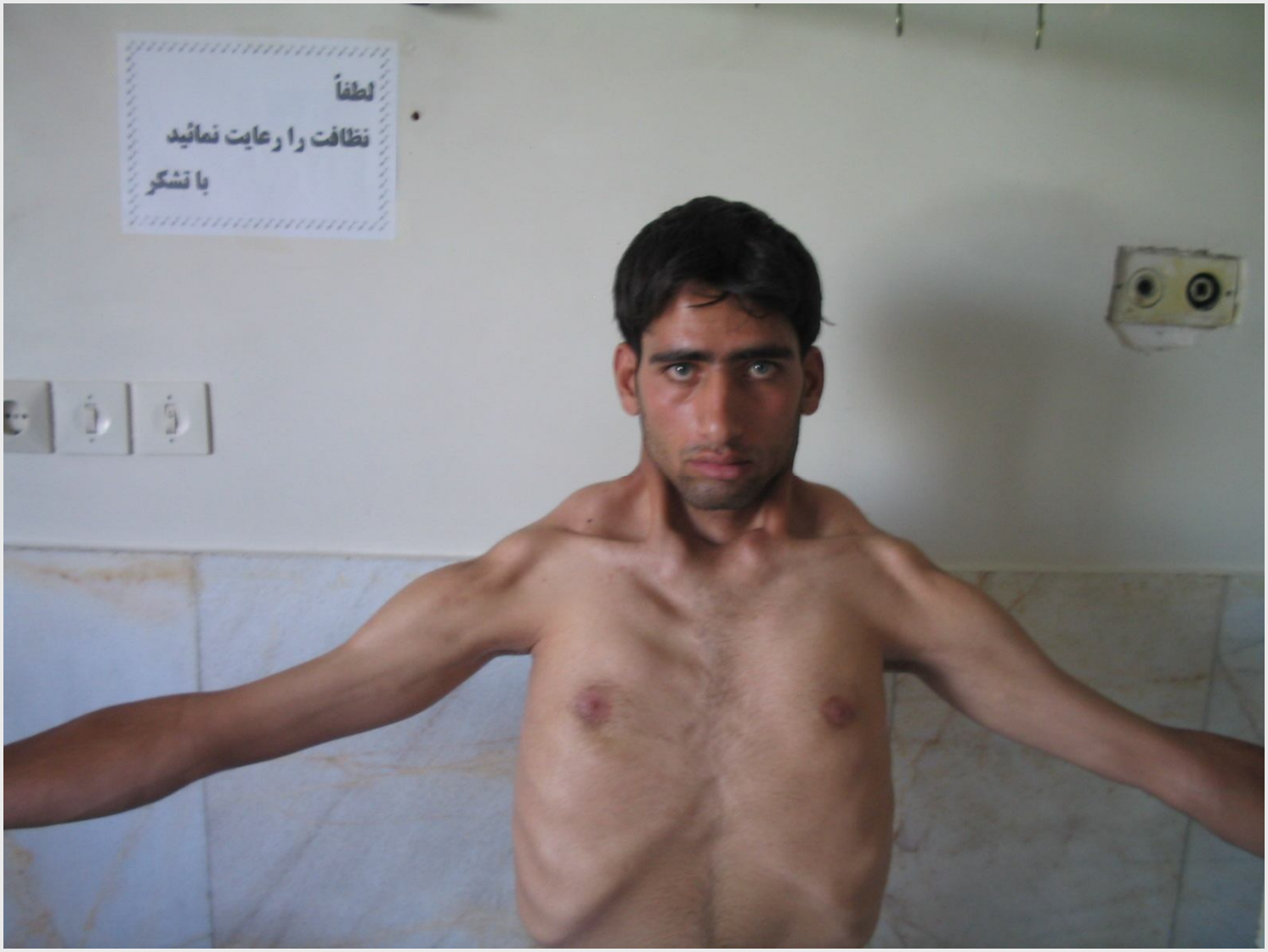
Bouche de tapir sign



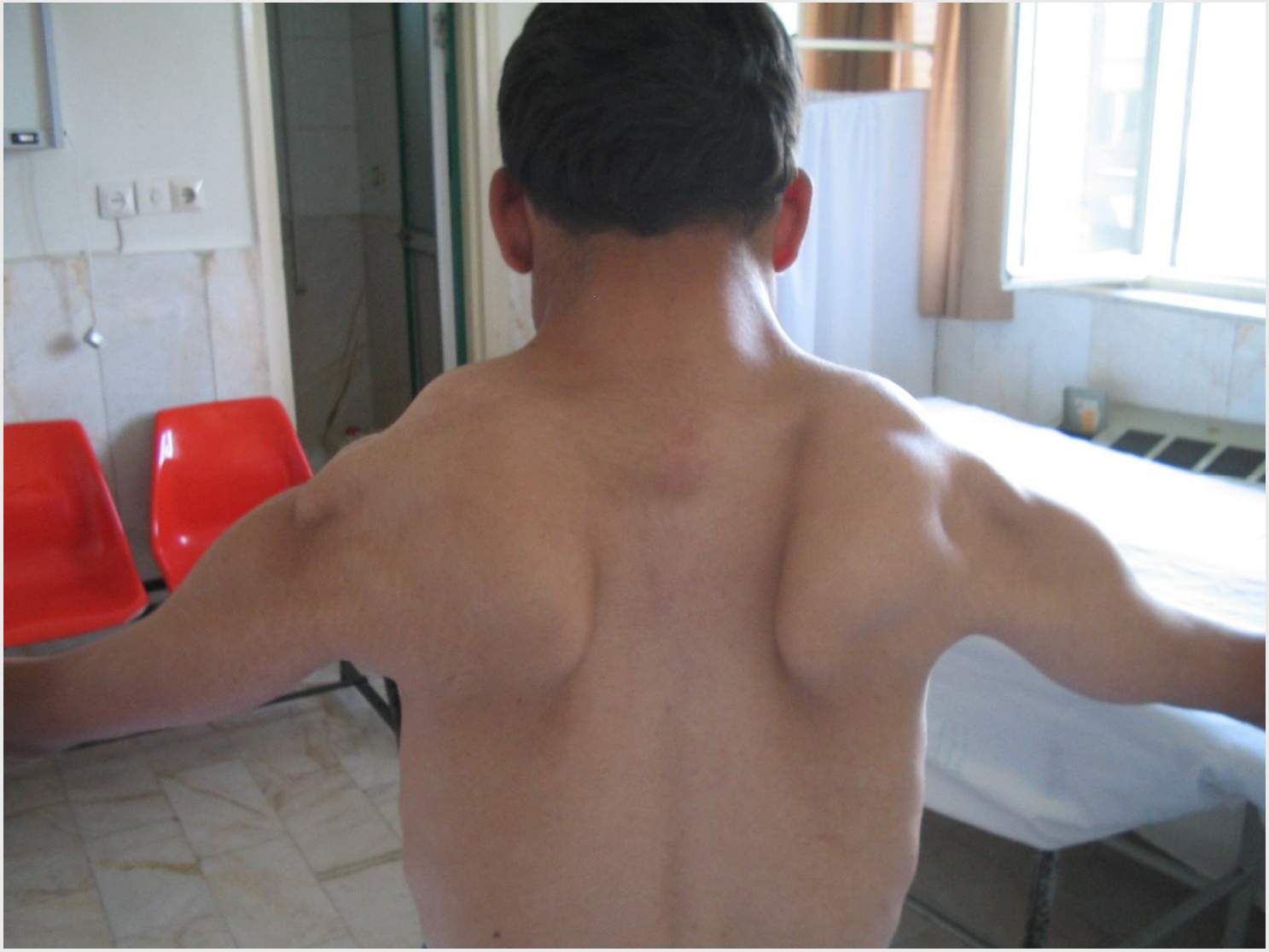
Popeye sign



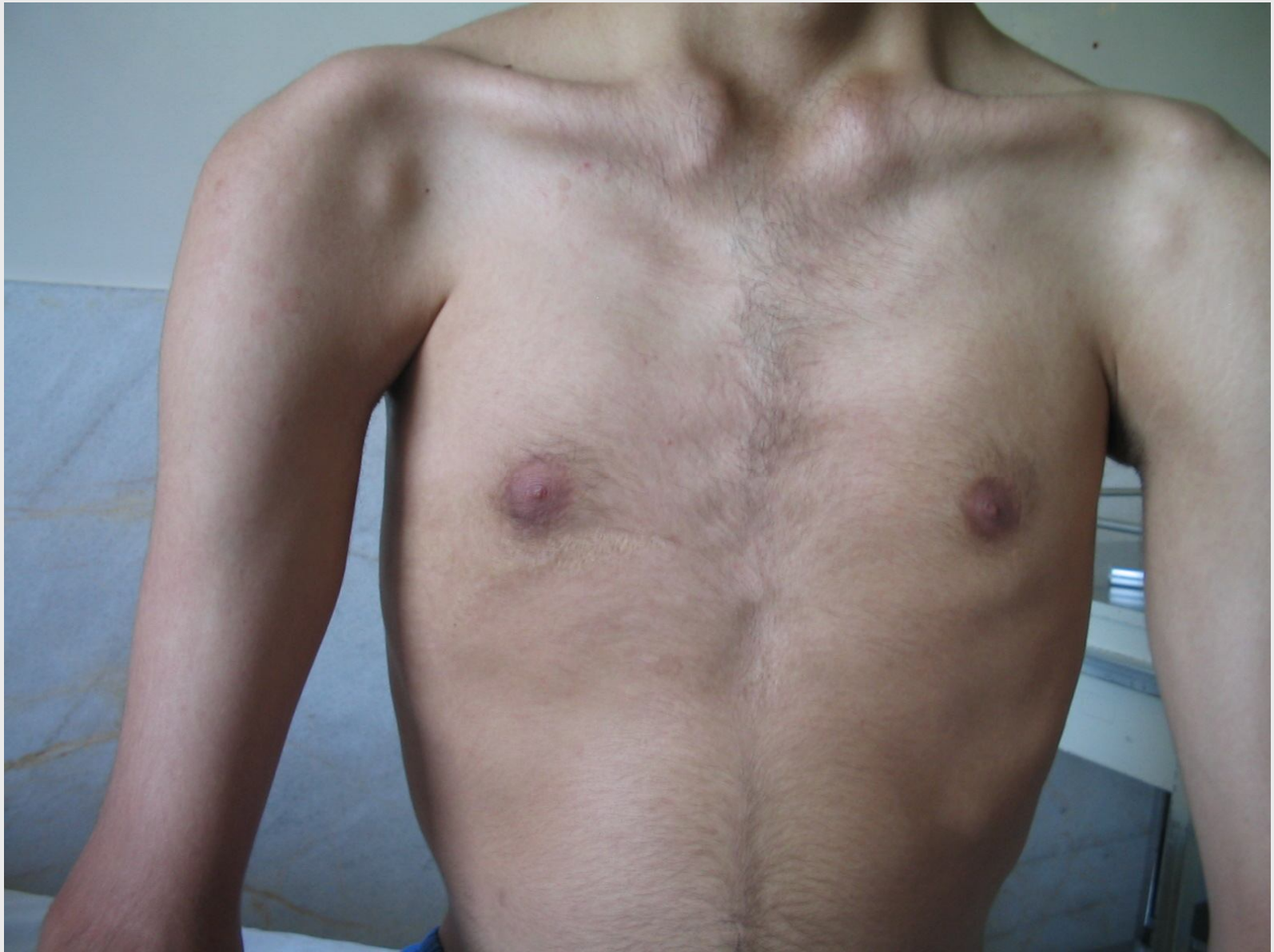
Trapezius hump



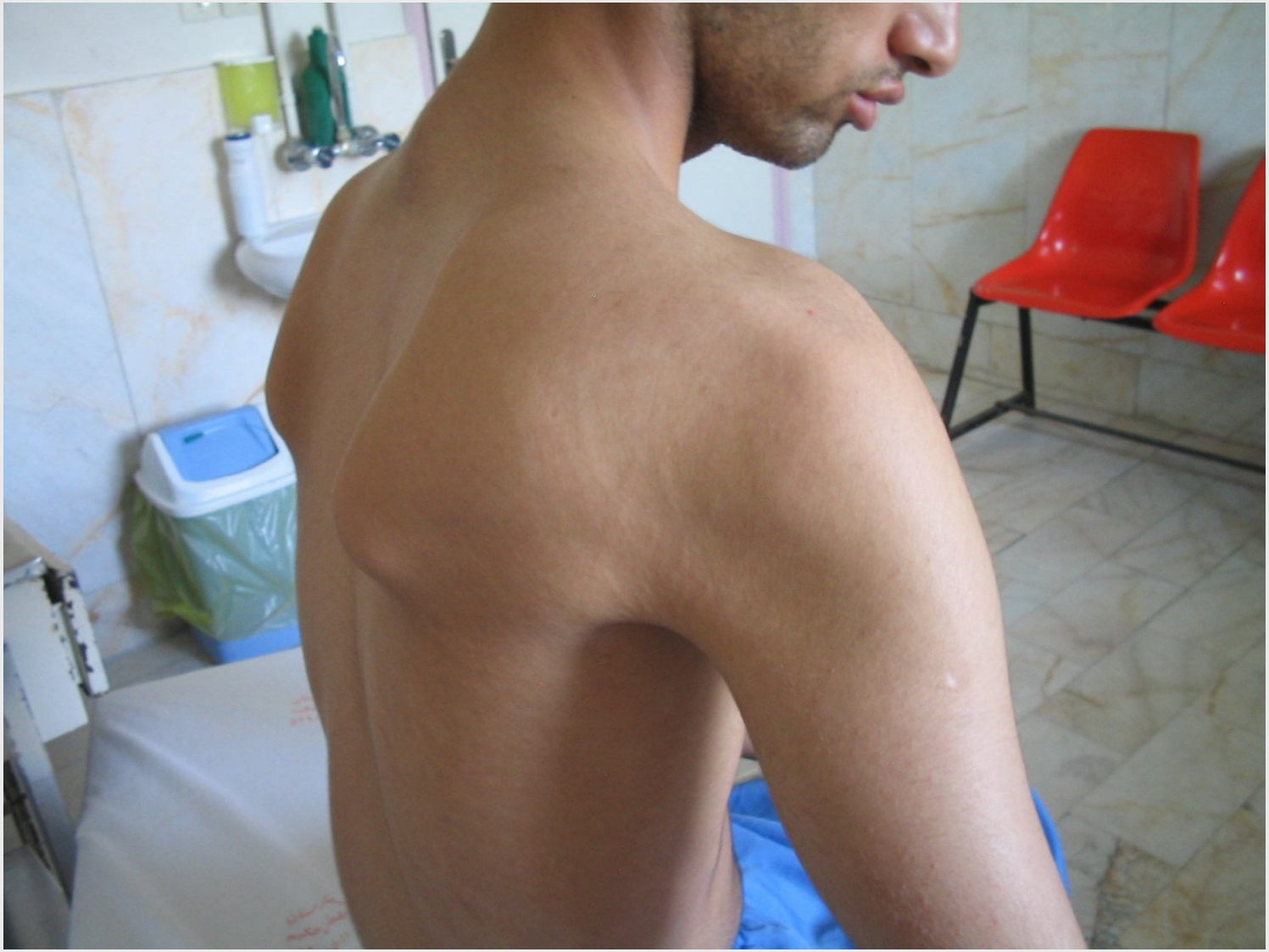
Winging of scapula



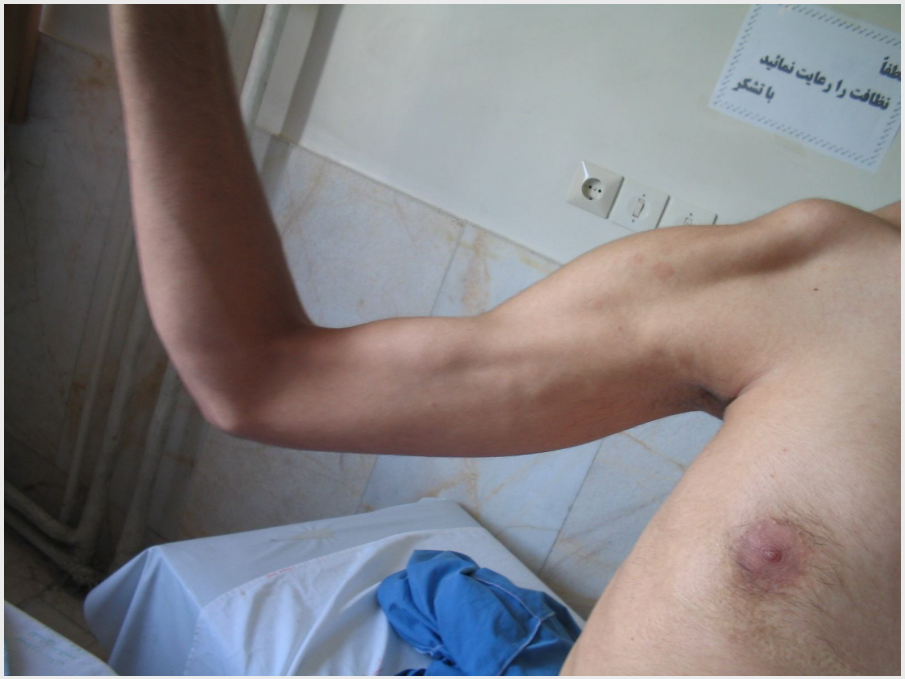
Muscle atrophy in proximal trunk

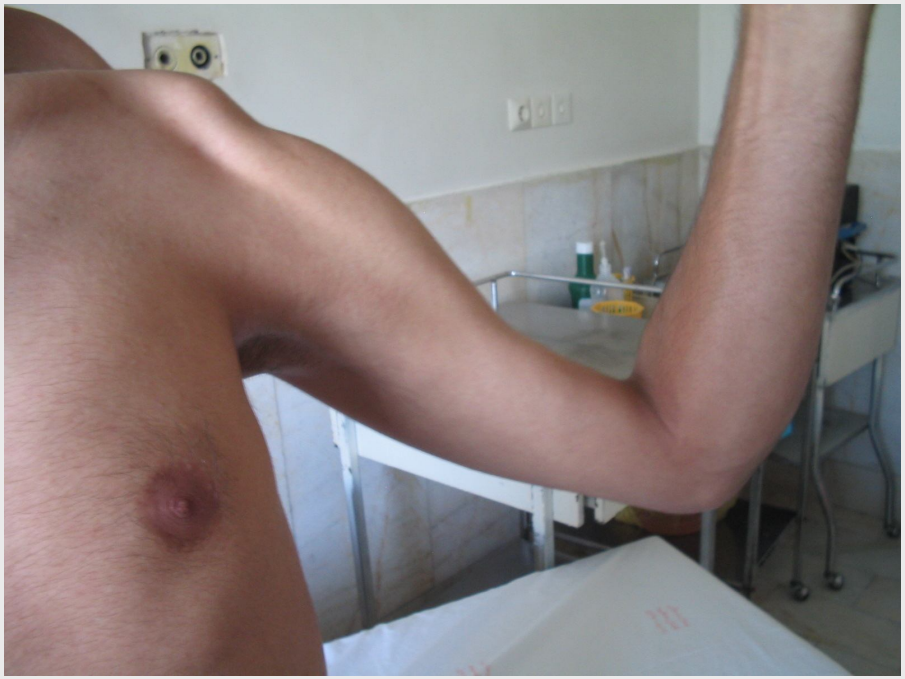


Muscle atrophy in shoulder girdle

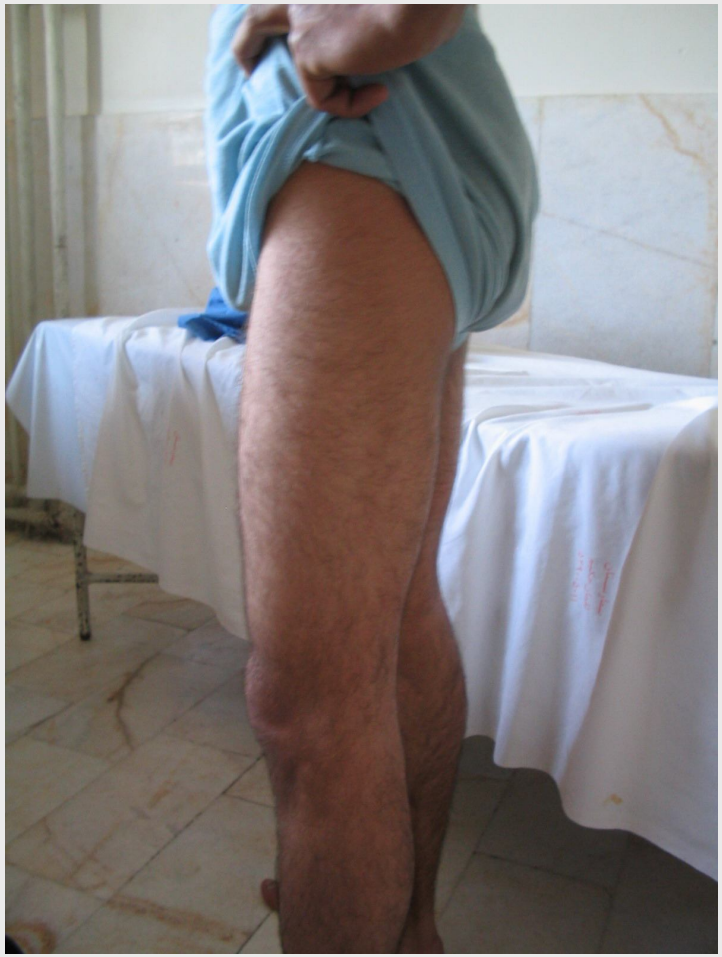


Atrophy in arm muscles

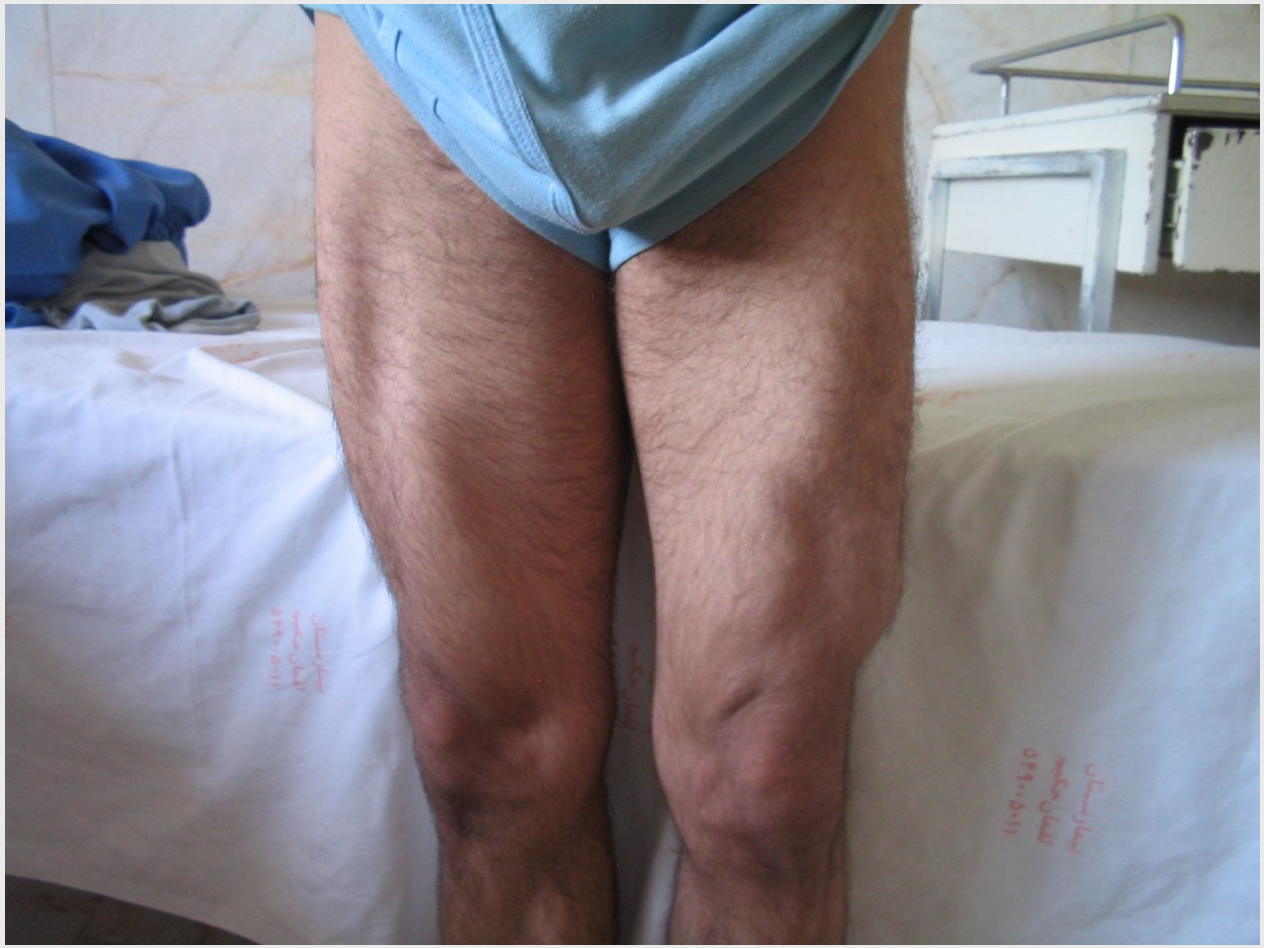




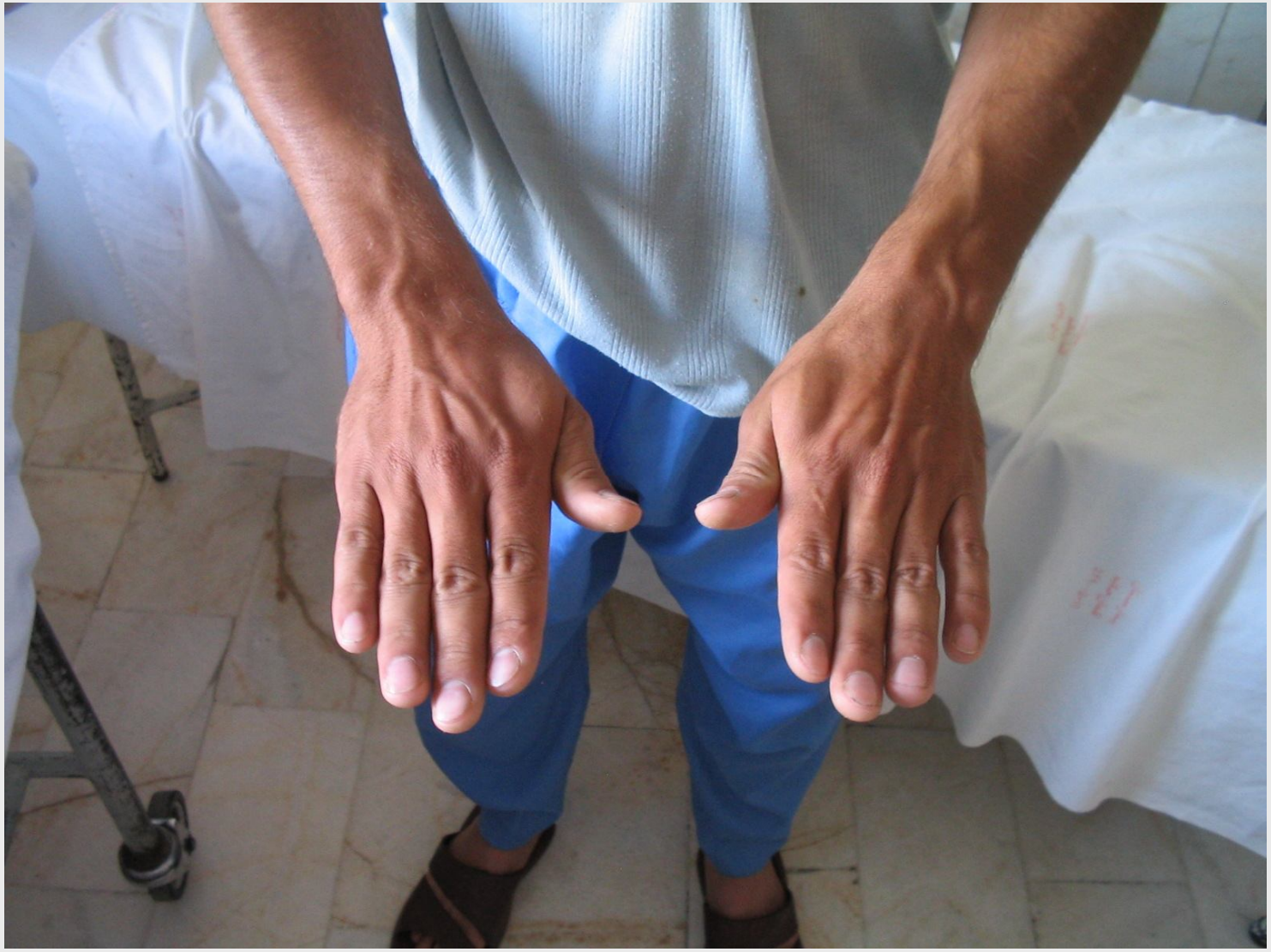
Mild atrophy in quadriceps



Mild atrophy in proximal thigh



Distal muscle sparing



localization

- UMN (Spaticity, Babinski's sign, Clonus, Hyperreflexia)
- LMN (flaccidity, areflexia, fasciculation, generalized atrophy)
- Radicuoplexopathy and neuropathy (pain, paresthesia, sensory loss, localized weakness, reflex loss)
- NMJ (ophtalmoplegia, fluctuation no atrophy)
- Muscular (weakness, atrophy, preserved reflex, special site involvement)

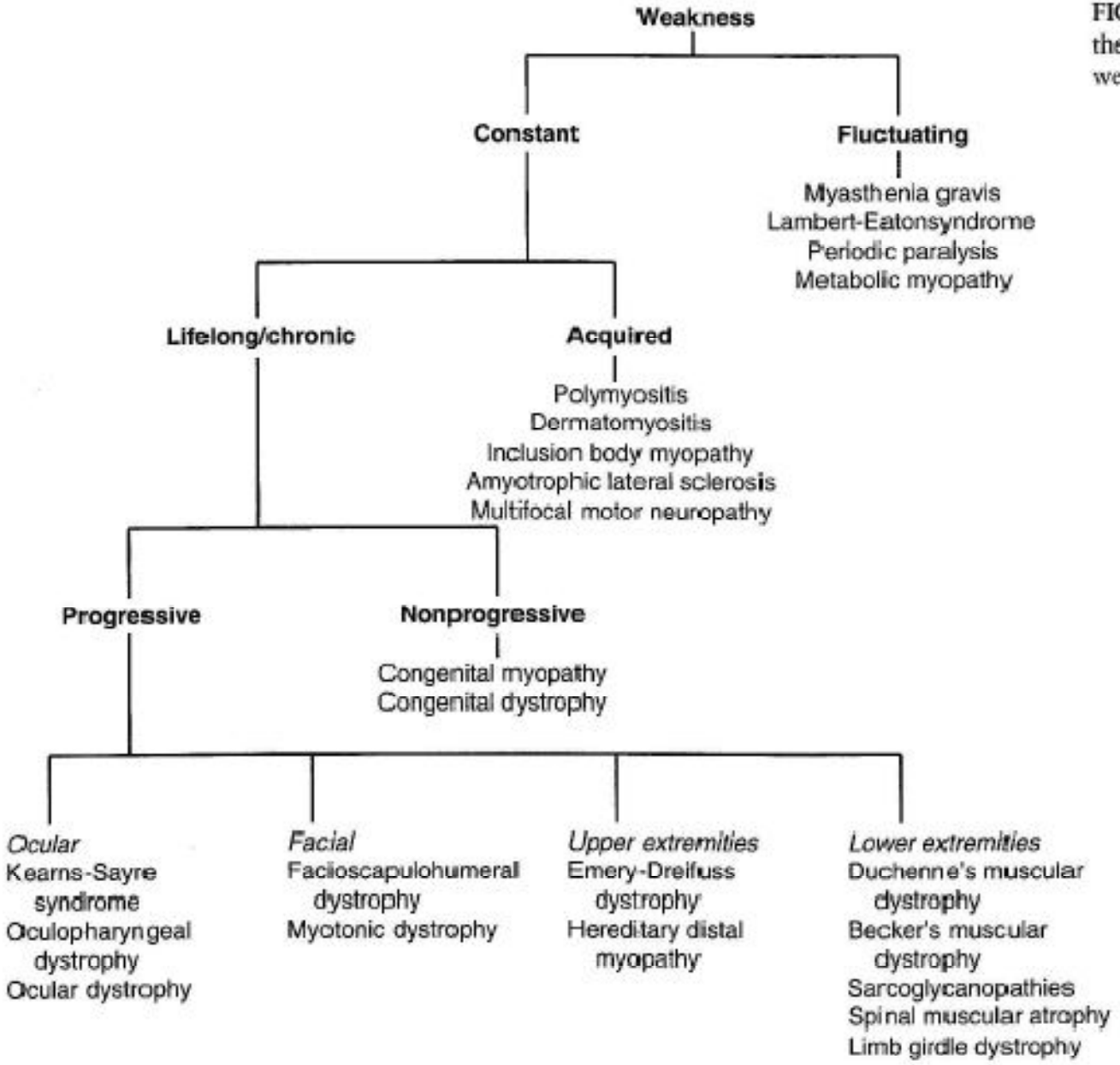


FIGURE 28.6 An algorithm for the approach to the patient with weakness.

Another ways to approach:
Approach to winging of scapula

Muscles involved in winging of scapula

- Trapezius (XI CN palsy or C3,4 radiculopathy).
- Rhomboid (Dorsal Scapular Nerve palsy or C4,5 radiculopathy).
- Levator of scapula (Dorsal Scapular Nerve palsy or C3,4,5 radiculopathy).
- Serratus anterior (Long Thoracic Nerve or C5-7 radiculopathy).
- Latissimus dorsi (Subscapular nerve or C5-8 radiculopathy).

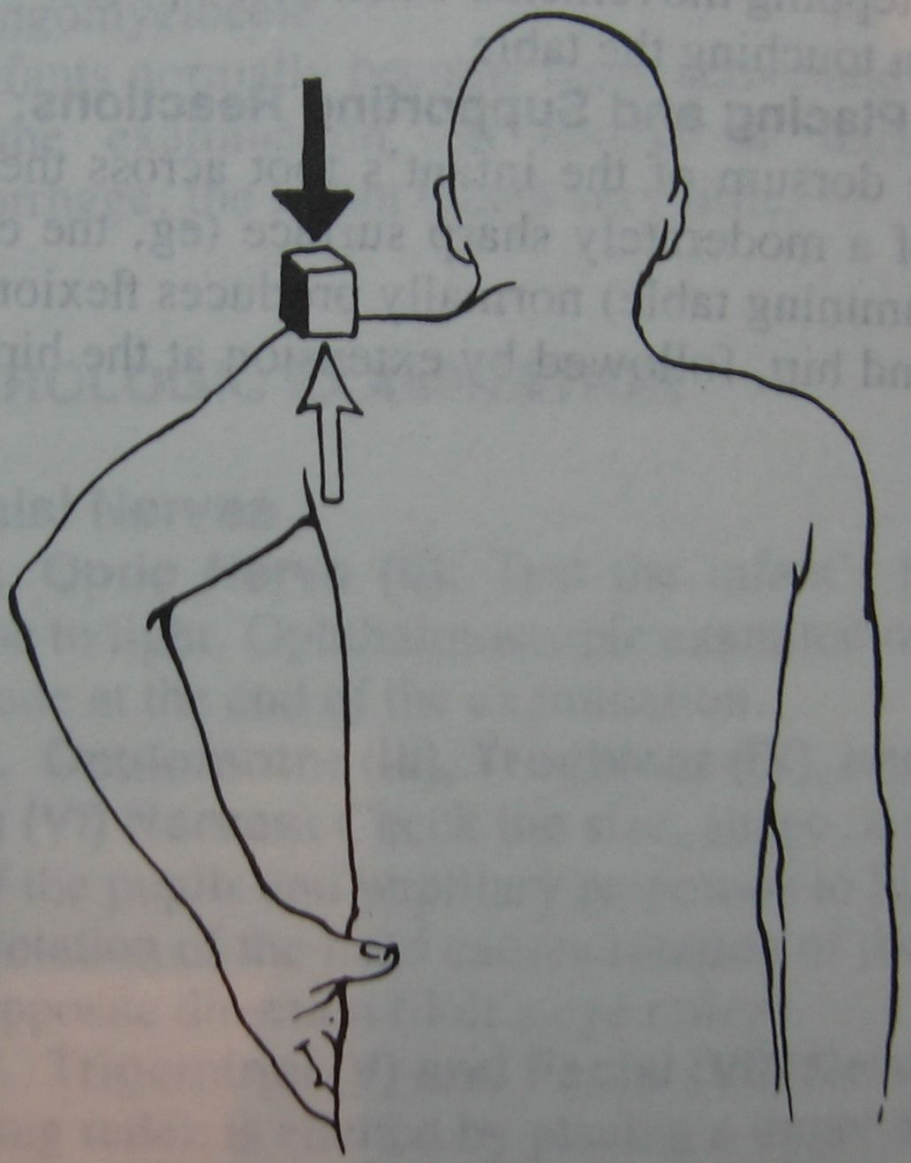


Figure 1. Trapezius, upper portion (C3, 4; spinal accessory nerve). The shoulder is elevated against resistance.

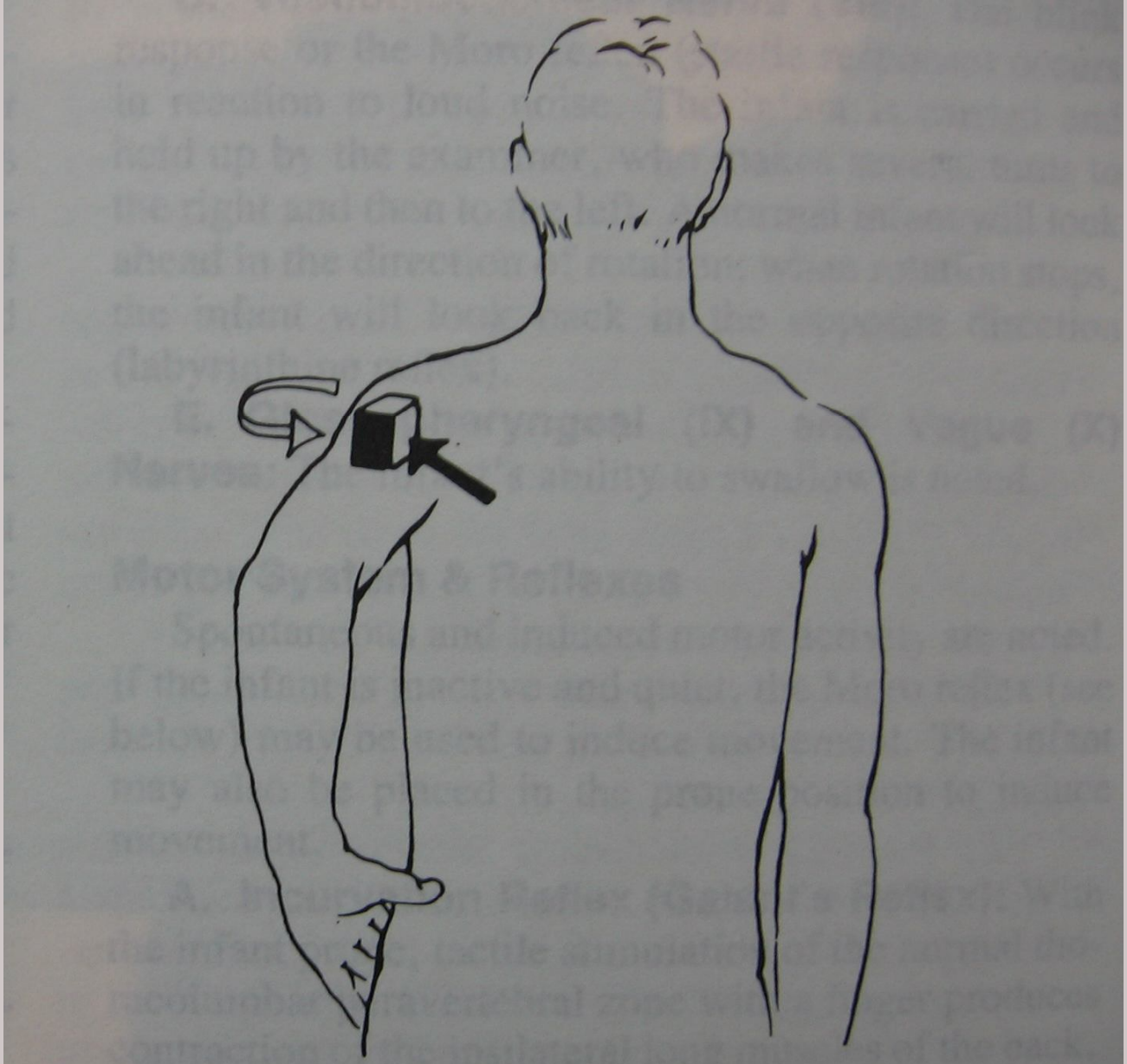


Figure 2. Trapezius, lower portion (C3, 4; spinal accessory nerve). The shoulder is thrust backward against resistance.

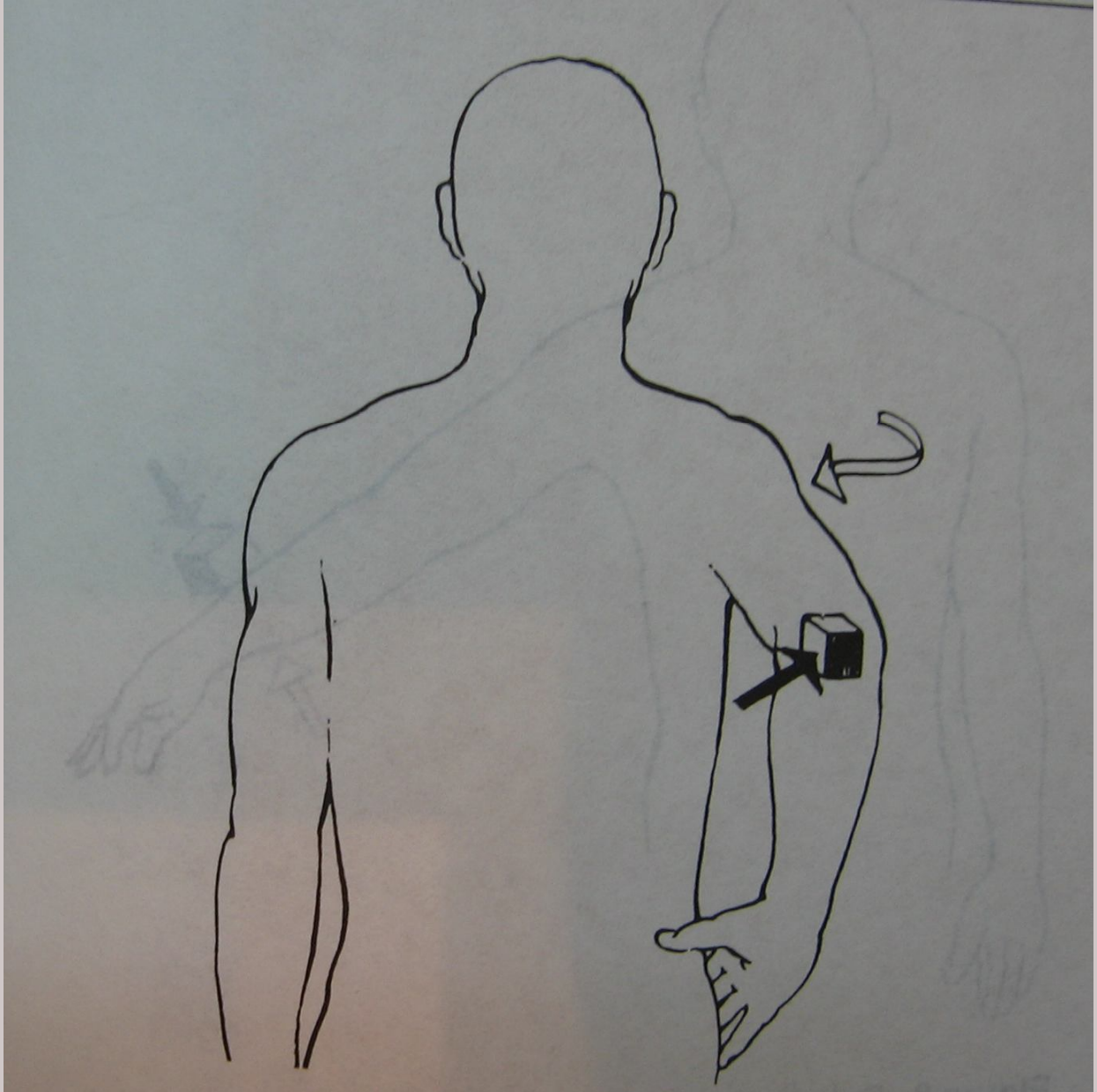


Figure 3. Rhomboids (C4, 5; dorsal scapular nerve). The shoulder is thrust backward against resistance.

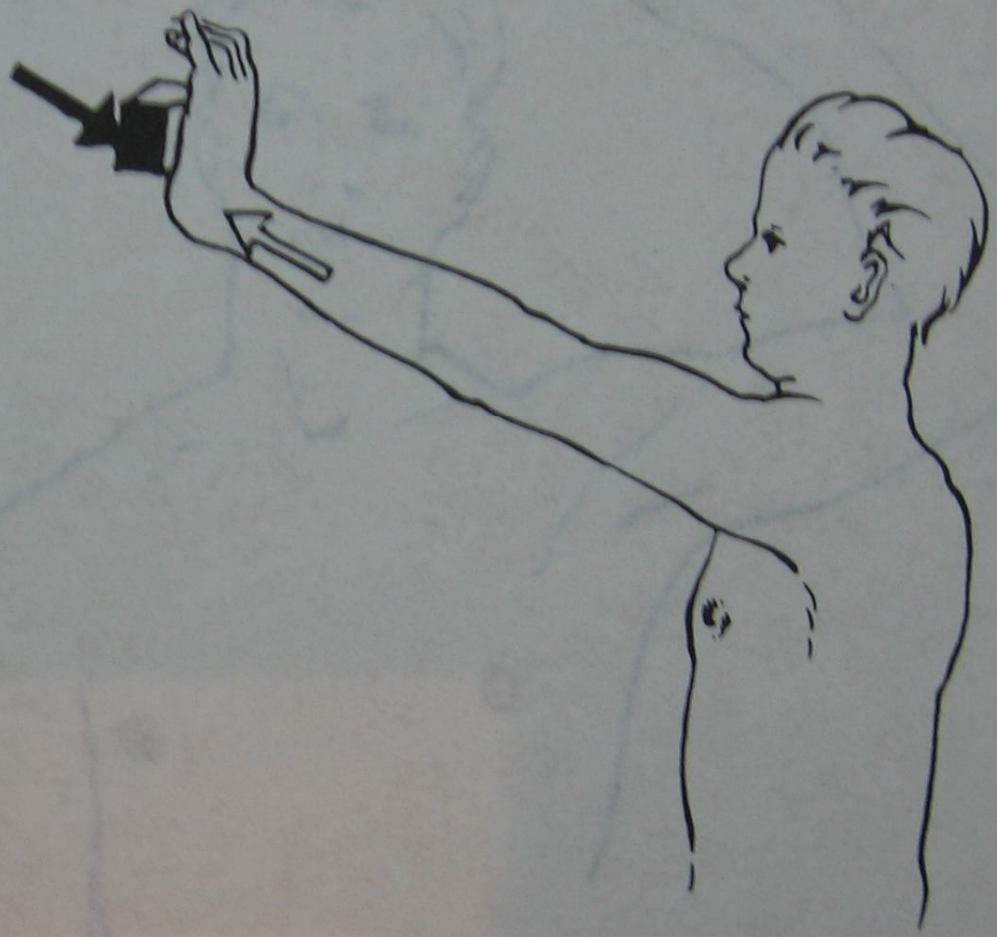


Figure 4. Serratus anterior (C5–7; long thoracic nerve). The patient pushes hard with outstretched arms; the inner edge of the scapula remains against the thoracic wall. (If the trapezius is weak, the inner edge may move from chest wall.)

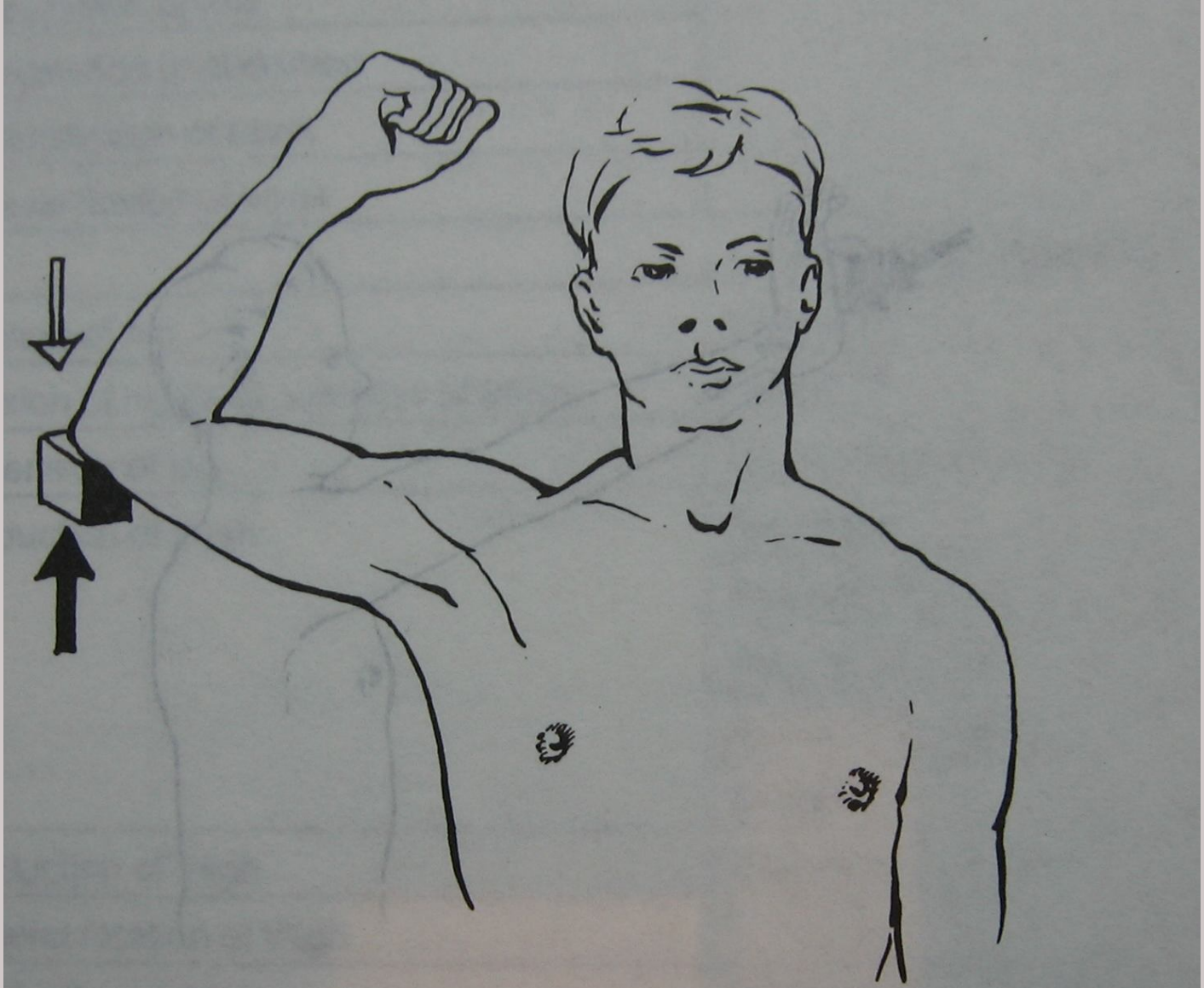


Figure 7. Latissimus dorsi (C5–8; subscapular nerve). The arm is adducted from a horizontal and lateral position against resistance.

Approach to winging of scapula

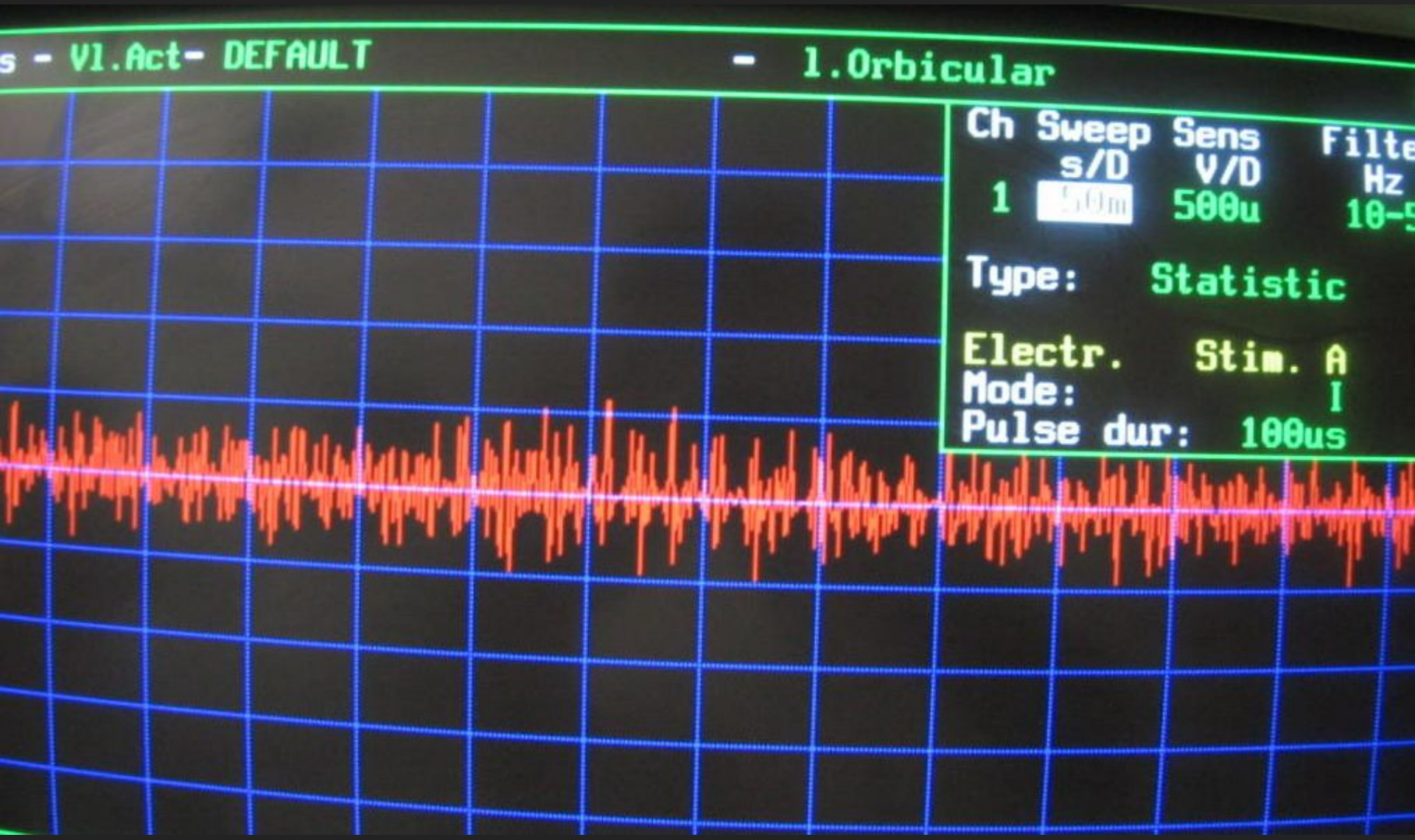
- **LGMD2A** (lower limb girdle atrophy and weakness and jutting backward of medial border of scapula).
- **XI CN Palsy** (winging with abduction but no hump and involvement of Trapezius and STM, and sparing of other muscles).
- **Long thoracic verve palsy and C7 radiculopathy** (C5,6,7 Ser. Ant.)[winging with forward elevation of arm but combination with other muscles involvement].
- **Dorsal scapular nerve and C5 radiculopathy** (weakness of the Rhomboid and levator of scapula muscles resulting in weakness of elevation and adduction of the medial border of the shoulder blade.) [winging with forward elevation of arm but sparing of other muscles].
- **Subscapular nerve or C5-8 radiculopathy (weakness of** Latissimus dorsi resulting in weakness of adduction in lateral position**).**
- **Idiopathic brachial plexopathy.**
- **Persons with low muscle bulk.**
- **FSHD (Trapezius hump and Popeye sign).**

Routine Lab test

- CPK 661
- LDH 491
- ESR 8
- AST 65
- AST 22
- CRP NEG
- VDRL NEG

EMG-NCV FINDING

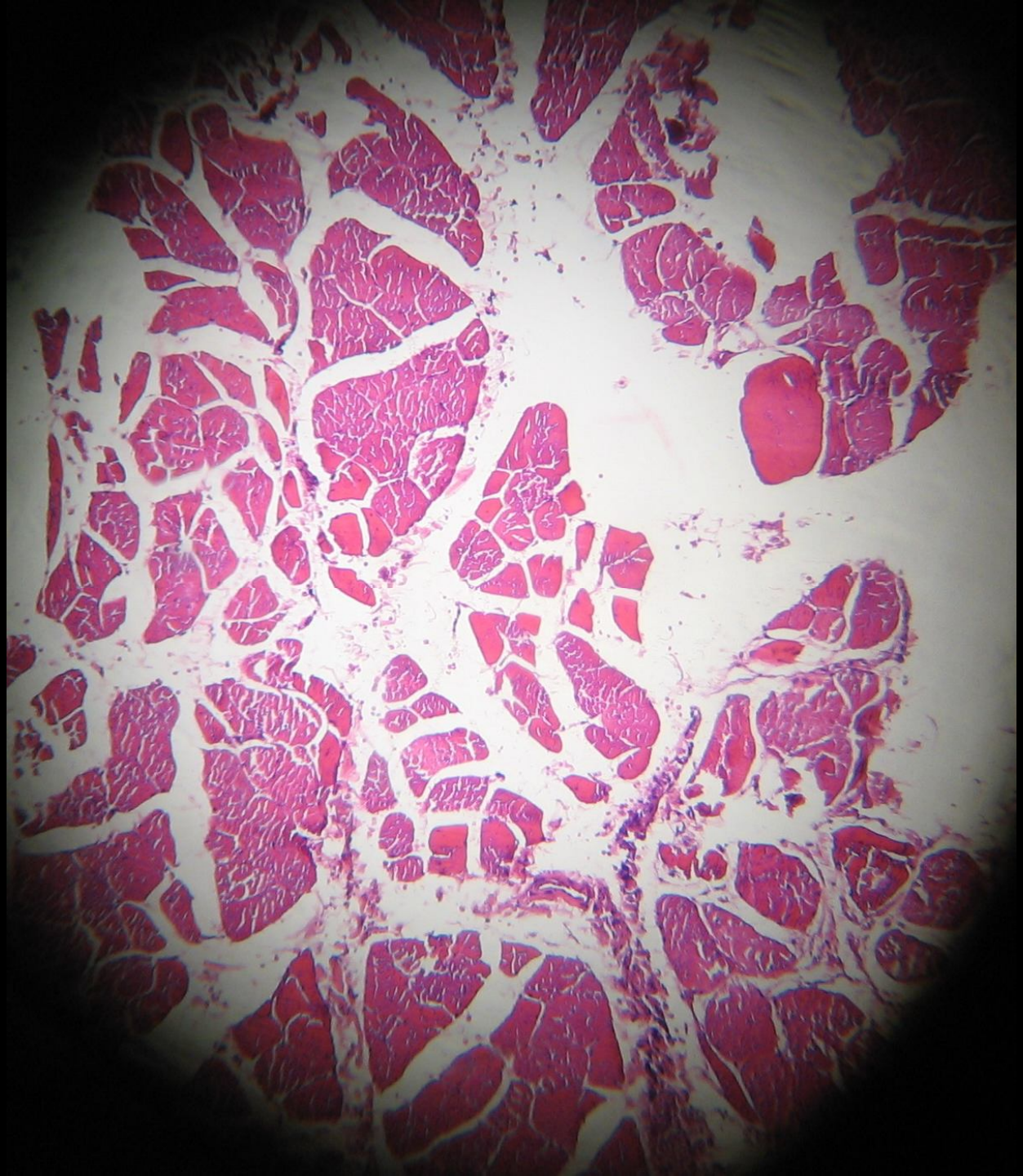
- NCV STUDY WAS NORMAL.
- EMG: MYOPATHIC CHANGES (POLYPHASIC MOTOR UNIT WITH REDUCED AMPLITUDE).

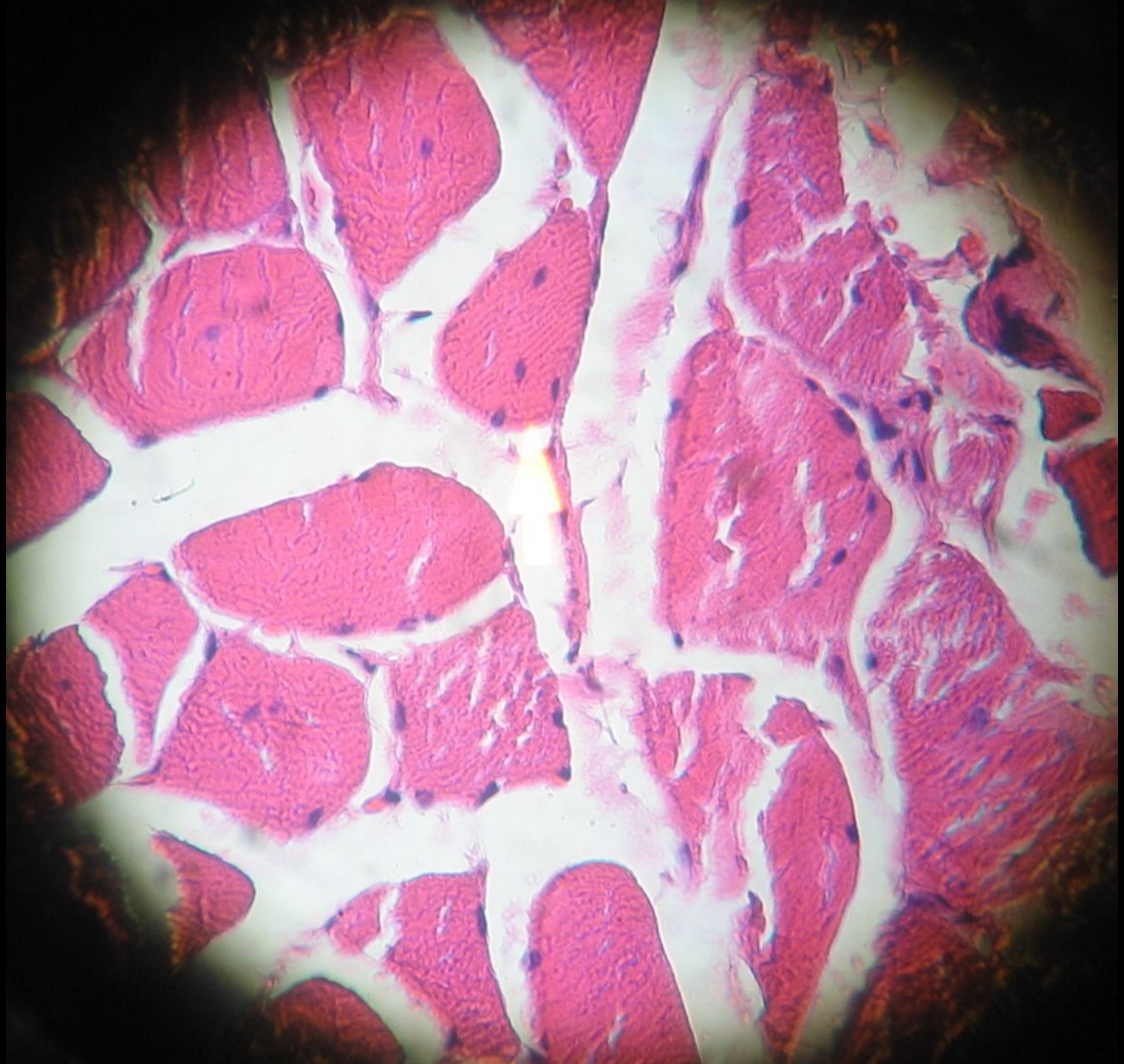

s - V1.Act- DEFAULT
- 1.Orbicular
Ch Sweep Sens Filte
s/D V/D Hz
1 50m 500u 10-5
Type: Statistic
Electr. Stim. A
Mode: I
Pulse dur: 100us

MUSCLE BIOPSY AND PATHOLOGICAL FINDING

- Fiber size variability.
- Fiber degeneration and regeneration.
- Fiber splitting.
- Hyaline changes.
- Central nucleus.
- Mild cellular inflammatory response.
- Endomesial and perimesial fibrosis.

These findings are in favor of muscular dystrophy.





ماکروسکپی :یک قطعه بافت عضله قهوه ای رنگ بابعاد ۲x۱x/۳ سانتیمتر باقطعات کوچکی ازبافت چربی اطراف چسبیده به آن میباشد

میکروسکپی :درمیزبیبنی ازبافت عضله سقط فیبرهای عضلانی باسایزهای متفاوت توکلئار-سنترالیزیشن وتغییرات دژنراتیو ورژنراتیو فیبرهامشهوده است درجائی از فیبروزاندوسیژیال وبوی میزیال دررنگ آمیزی های اختصاصی بچشم میخورد-

تشخیص :

DX: Deltoid muscle biopsy:
Non inflammatory myopathic disorder compatible with muscular dystrophy .

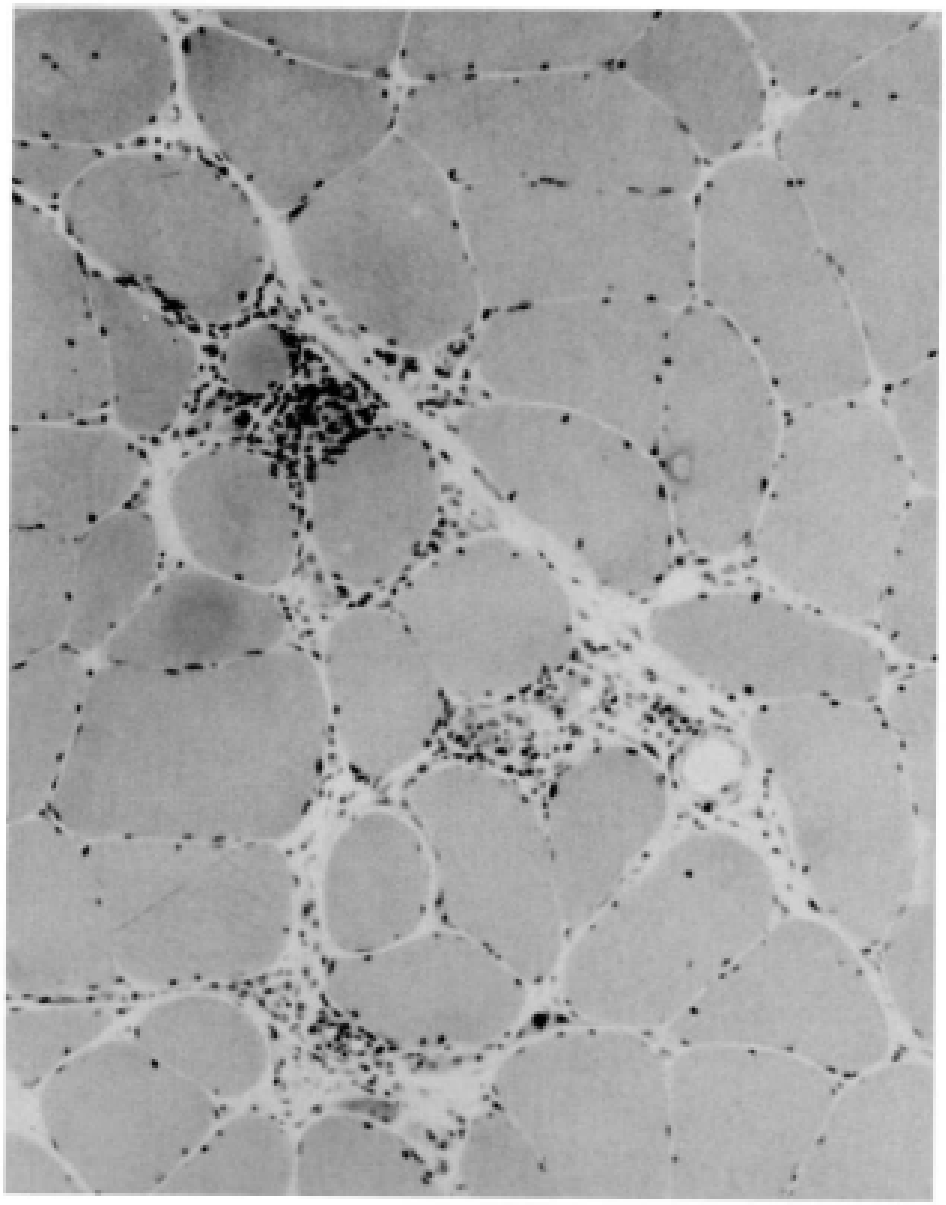


FIGURE 85.18 Facioscapulohumeral dystrophy. Cellular inflammatory responses are noted in the biopsies from many patients with this illness. These are more often associated with necrotic fibers than with blood vessels. (Hematoxylin-eosin stain.)

FINAL DIAGNOSIS

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

- FSHD is inherited as an autosomal dominant trait. Its prevalence is 1-2 per 100,000 population (incidence rate 1 in 20,000).
- The responsible gene has been localized to chromosome 4q35 in many, but not all, families.
- The most common time of presentation is during adolescence but the age of onset is from infancy to middle age.

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

- The severity of FSHD varies even within the same family.
- Some patients may have only mild facial weakness whereas others have total paralysis of the face and severe weakness of other muscles causing wheelchair confinement during childhood.

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

There are two distinctive pattern of muscular weakness of face, scapular stabilizer, proximal arm, and peroneal muscles.

- **The first is gradually descending AD form.**
- **The second is a jump form in which the progressive weakness jumps from the upper body to the peroneal muscles.**

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

- The initial weakness typically affect the facial muscles, specially orbicularis oculis and oris.
- The masseter, temporalis, extraocular, and pharyngeal muscle are usually unnaffected.
- Shoulder weakness is presenting symptoms in %85 of symptomatic patients.

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

- Involvement of the scapular fixator muscles, the latissimus dorsi, trapezius, rhomboids, and serratus anterior muscles causes a winging of scapula, a highly characteristic sign.
- The scapula is placed more laterally than the normal. It moves upwards in shoulder abduction.
- The deltoid muscle typically not affected.

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY

- **The most severe form of FSHD occurs in infancy. Such babies have severe weakness.**
- **The face has no movement and remains passive and expressionless.**
- **Affected children may sleep with the sclera of the eyes showing through partially opened lids (normal cornea).**

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

- Weakness of the limbs, although it conforms to the general pattern of FSHD, is so severe that the ability to walk is lost by 9 or 10 years of age.
- A striking feature is the extreme lumbosacral lordosis seen when the child walks or stands. This initially disappears on sitting, indicating that a compensatory mechanism is used to maintain balance.
- Deafness and Coats' disease, an oxidative vascular degeneration of the retina, are associated abnormalities.

Diagnosis

- The diagnosis can be established reliably by DNA studies.
- The serum CK concentration is elevated, muscle histological preparations shows myopathic features, sometimes with scattered inflammatory foci.
- The EMG shows myopathic potentials.

Treatment

- Prednisone has been tried in some patients, but the results are generally not beneficial, unless there is significant inflammation in the muscle biopsy.
- Orthopedic and supportive measures.