Neuromuskulární poruchy u thyreopatií

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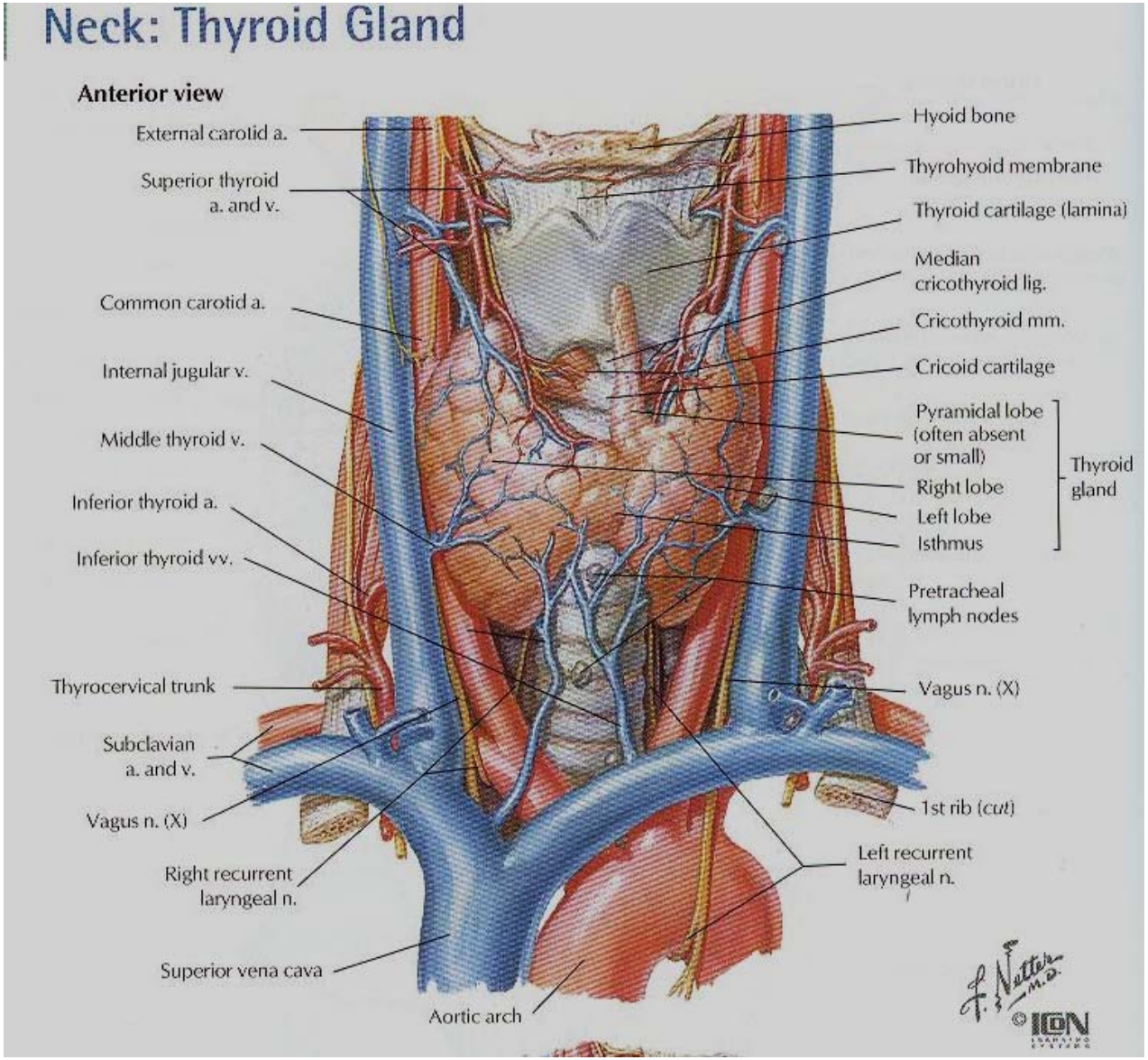
Anatomie


Neck: Thyroid Gland
Anterior view
External carotid a.
Superior thyroid a. and v.
Common carotid a.
Internal jugular v.
Middle thyroid v.
Inferior thyroid a.
Inferior thyroid vv.
Thyrocervical trunk
Subclavian a. and v.
Vagus n. (X)
Right recurrent laryngeal n.
Superior vena cava
Aortic arch
Hyoid bone
Thyrohyoid membrane
Thyroid cartilage (lamina)
Median cricothyroid lig.
Cricothyroid mm.
Cricoid cartilage
Pyramidal lobe (often absent or small)
Right lobe
Left lobe
Isthmus
Thyroid gland
Pretracheal lymph nodes
Vagus n. (X)
1st rib (cut)
Left recurrent laryngeal n.
F. Netter M.D.
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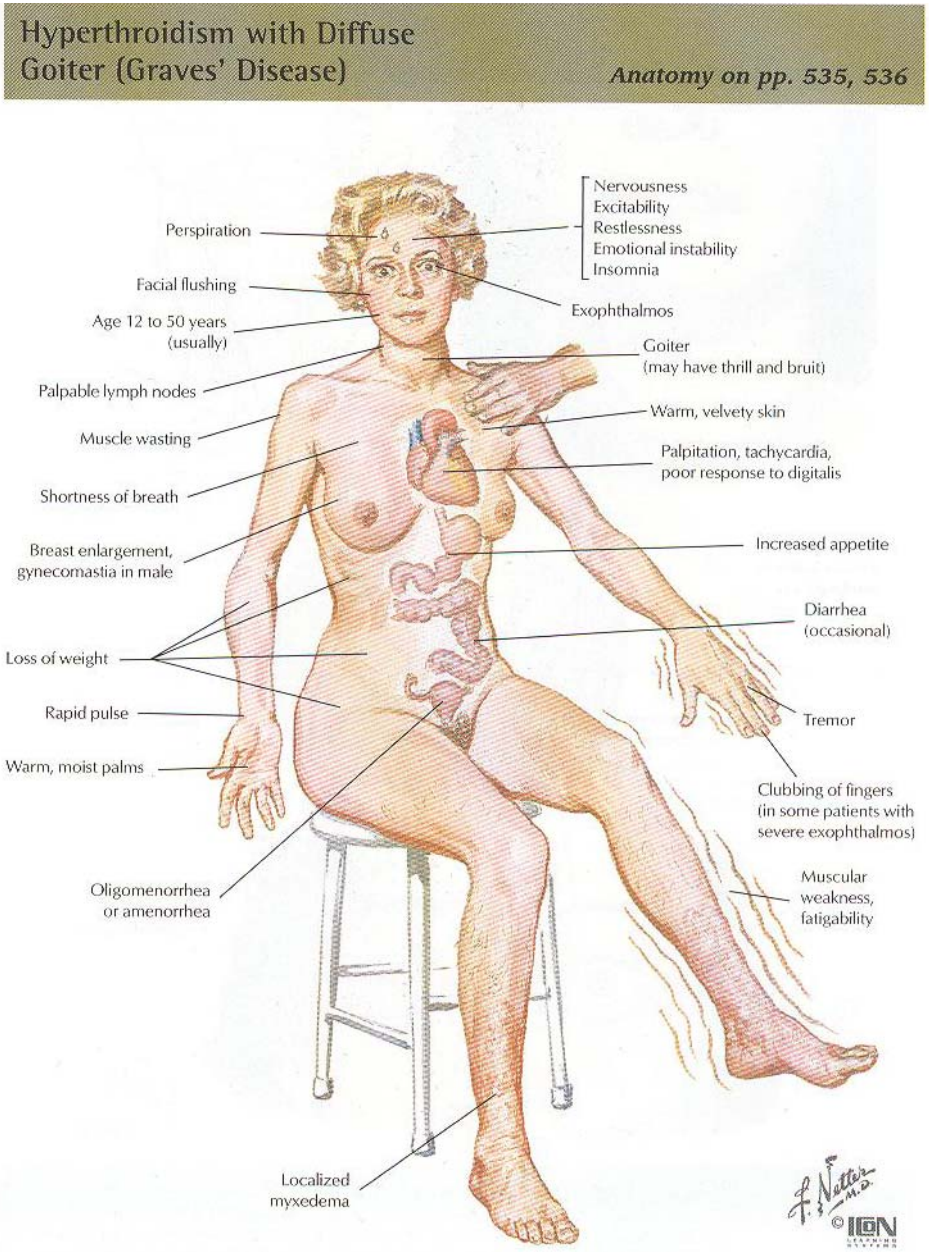
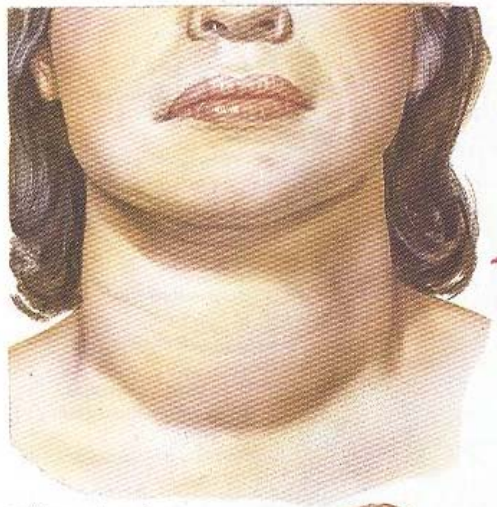
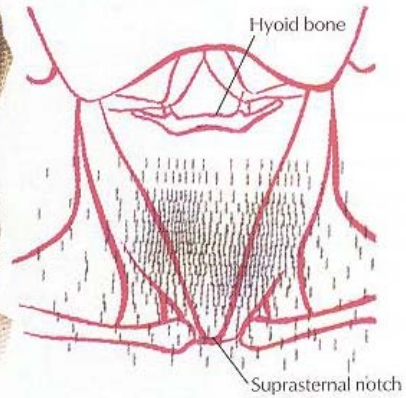
Hyperfunkce

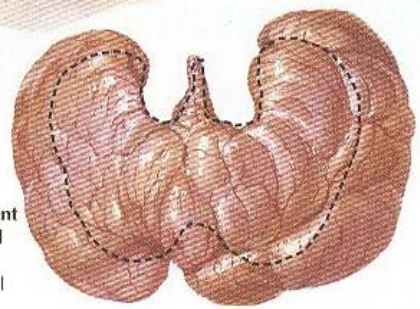






Scintigram

Diffuse goiter of moderate size



Diffuse enlargement and engorgement of thyroid gland (broken line indicates normal size of gland)

Moderately severe exophthalmos

1.9% žen a 0.19% mužů

NM poruchy u hyperthyreózy

Thyreotoxická myopatie

Thyreotoxická periodická paralýza

Endokrinní (Gravesova) orbitopatie (oftalmopatie)

Distální převážně senzitivní axonální neuropatie

Thyreotoxická myopatie

- Frekvence stejná u mužů i žen
- Závažnost koreluje s délkou hyperthyreózy
- Symptomy za 1-3 měsíce od manifestace

- Vývoj obvykle pozvolný (měsíce)
- Vzácně akutní nástup a průběh thyreotoxikózy vede k rhabdomyolýze a myoglobinúrii

Thyreotoxická myopatie

- Proximální svalová slabost asi 5%, převážně únava 75% (většinou DK)
- Distální slabost cca 20%
- Časté myalgie, krampy a fascikulace
- Reflexy bývají normální, u 25% vyšší

- CK normální - nekoreluje se svalovou slabostí, vysoké fT3 + fT4, nízké TSH
- EMG – abnormní u sval.slabosti, bez svalové slabosti abnormní cca u 1/3 nemocných
- **Svalová biopsie – nespecifická** - atrofie vláken 1 a 2.typu a zmnožená vnitřní jádra

Kazuistika perzistentní thyreotoxické myopatie

Ve 49 letech ataka

hyperthyreózy s oftalmopatií

pro riziko amaurózy ⟹

1998 totální strumektomie +

léčba ^{131}I a následně

6 měsíců bez substituce ⟹

vývoj slabosti pelvifemorálních

svalů (dřepy, kolo)

2010 stále trvá svalová slabost

Thyreotoxická periodická paralýza

- Ztráta excitability sarkolemy v důsledku inaktivace Na^+ kanálů
- Asiaté 10 %, běloši 10x méně (Satoyoshi et al.1998)
- Častěji muži 6:1

- **Typické epizodické ataky svalové slabosti v trvání hodin až dnů**
- Generalizované formy nebo sval.skupiny
- Provokace – alkohol, sladkosti, chlad
- CK – vysoká, K^+ 1.1-3.4 mmol/l, NCV – ↓ CMAP
- Dif.dg.hypokalemická PP

Gravesova orbitopatie (oftalmopatie)

- Zvýšený obsah orbit (voda, glykoproteiny, zánětlivá celulizace v retro-okulární pojivové tkání)
- Proliferace fibroblastů v EO svalech → ztluštění
- 5% osob s hyperthyreózou vyvine oftalmopatii
- Klinika má 7 stupňů (0-6) (J.Bednařík : Nemoci koster.svalstva,2004)
- Ohrožení amaurózou 6%
- Cave : manifestace nebo progrese i po strumektomii
- Dg: sono + MR orbit – ztluštění EO svalů

Asociace s myasthenia gravis

- potvrzená asociace MG + thyreopatie ve 13%

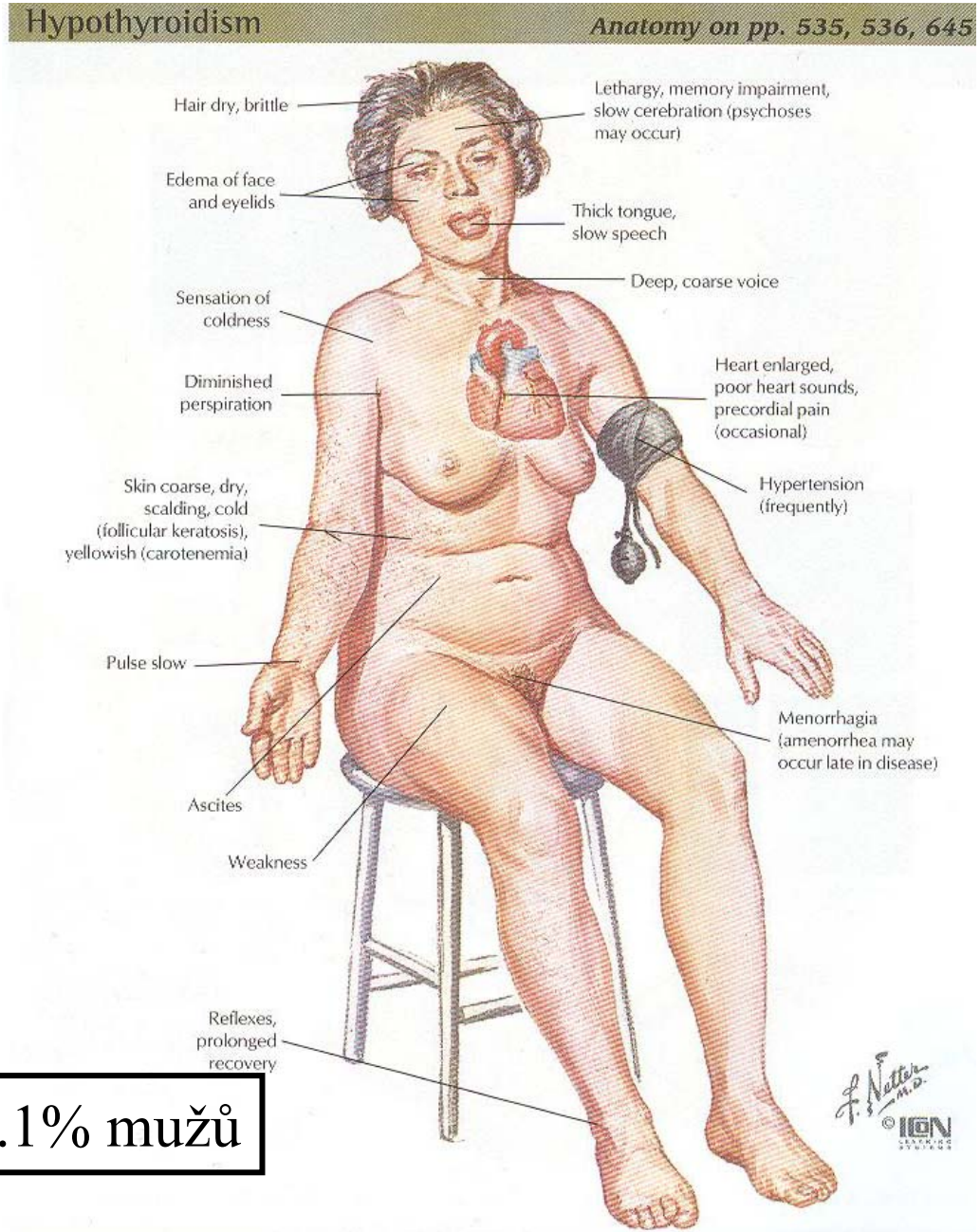
- 5.7% trpí hyperthyreózou
- 5.3% jsou hypothyreoidní
- 2.1% non toxická struma

(J.Bednařík : Nemoci KS, Triton 2004, Katiriji et al. 2007)

Postižení PNS u hyperthyreózy

- 20% pacientů vykazuje **axonální** senzitivní a motorickou neuropatie
- klinicky dominují **senzitivní** neuropatické symptomy
- úžinové syndromy velmi vzácné

Hypofunkce



1.4% žen a 0.1% mužů

Hypothyreoidní myopatie

- krampy, bolesti a tuhost svalů – 75%
- svalová slabost – 30% - včetně respiračních svalů
- korelace s délkou a tíží hypothyreózy

- CK je zvýšená až 10x při svalové slabosti
- EMG : normální nebo úzké MUPs
- Sval.biopsie – atrofie a někdy nekróza sval.vláken

Postižení PNS u hypothyreózy

- úžinové léze – CTS u 25% pacientů

- polyneuropatie – **senzitivní axonální** u **19% pacientů**

NM poruchy u hypothyreózy

79% pacientů NM poruchy

38% svalová slabost jednoho či více svalů

42% axonální senzitivně motorická neuropatie

<u>29% sy canalis carpi</u>

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J Neurol Neurosurg Psychiatry 2000;68:750-755 doi:10.1136/jnnp.68.6.750

Paper

Neuromuscular findings in thyroid dysfunction: a prospective clinical and electrodiagnostic study

Ruurd F Duyff[a], Joan Van den Bosch[c], D Martin Laman[b], Bert-Jan Potter van Loon[c], Wim H J P Linssen[a]

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Abstract

OBJECTIVES To evaluate neuromuscular signs and symptoms in patients with newly diagnosed hypothyroidism and hyperthyroidism.

METHODS A prospective cohort study was performed in adult patients with newly diagnosed thyroid dysfunction. Patients were evaluated clinically with hand held dynamometry and with electrodiagnosis. The clinical features of weakness and sensory signs and the biochemical data were evaluated during treatment.

RESULTS In hypothyroid patients 79% had neuromuscular complaints, 38% had clinical weakness (manual muscle strength testing) in one or more muscle groups, 42% had signs of sensorimotor axonal neuropathy, and 29% had carpal tunnel syndrome. Serum creatine kinase did not correlate with weakness. After 1 year of treatment 13% of the patients still had weakness. In hyperthyroid patients 67% had neuromuscular symptoms, 62% had clinical weakness in at least one muscle group that correlated with FT4 concentrations, but not with serum CK. Nineteen per cent of the patients had sensory-motor axonal neuropathy and 0% had carpal tunnel syndrome. The neuromuscular signs developed rapidly, early in the course of the disorder and were severe, but resolved rapidly and completely during treatment (average time 3.6 months).

CONCLUSIONS Neuromuscular symptoms and signs were present in most patients. About 40% of the hypothyroid patients and 20% of the hyperthyroid patients had predominantly sensory signs of a sensorimotor axonal neuropathy early in the course of thyroid disease. Weakness in hyperthyroidism evolved rapidly at an early stage of the disorder and resolved completely during treatment, suggesting a functional muscle disorder. Hand held dynamometry is sensitive for the detection of weakness and for the clinical evaluation of treatment effects. Weakness in hypothyroidism is more difficult to treat, suggesting myopathy.

JNNP, 2000

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67% pacientů NM poruchy

62% únava jednoho a více svalů, 25% slabost

19% axonální senzitivně motorická neuropatie

0% sy canalis carpi

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Duyff RF: JNNP, 2000

Terapie

Euthyreoidní stav – racionální výstup léčby

Thyreotoxická myopatie – slabost respiračních svalů ⇒ β blokátory (Wang a Poh, 1986)

Thyreotoxická PP ⇒ přívod K^+

Orbitopatie/oftalmopatie ⇒ prednisolon + cyklosporin s 50% efektem

dekompresní operace


Děkuji za pozornost