

Dysautonomias

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Structure of autonomic nervous system

Functions of ANS

- influences all body processes, which are not directly related to posturation and movement
- processes can be little influenced by will and unconscious
- ANS does not command rather modulates visceral functions

Modulation and control of

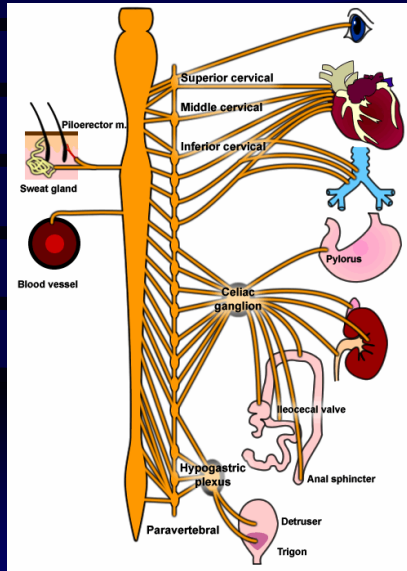


1. visceral organ functions (heart, GIT)
2. smooth muscles (vessels, airways, urogenital, piloerectores)
3. glandular secretion (napr. saliva, tears, mucus, sweat)
4. endocrine glands (nervous sy - endocrine sy. interaction)
5. immune system (nervous sy - immune sy. interaction)
6. metabolic processes (e.g. liver, muscles, fat tissue, etc.)
7. heat production (liver, muscles)
8. trophic functions (skin, connective tissue, etc.)

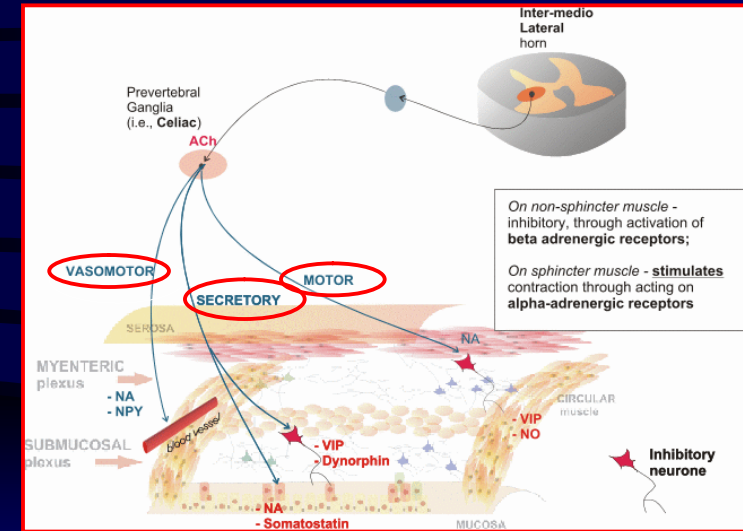
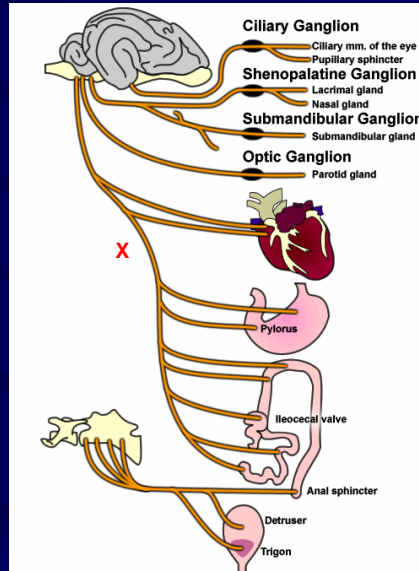
Peripheral components of ANS / 1

- **Sympathetic system** (cervical-thoracic-lumbar level)
- **Parasympathetic system** (cranial-sacral level) n. vagus
- ⊗ **Visceromotoric pathways** (interomotoric/ glandulomotoric)
- ⊗ **Viscerosensitive pathways** (interoceptive/ glanduloceptive)
 - **Preganglionic neurons**
(visceromotoric nuclei of cranial nerves, intermediolateral column)
 - **Postganglionic neurons**
(paravertebral ganglia, isolated ganglia close to organs)
Sympathetic: gg. cervicale, g. celiacum, g. mesenterici,
Parasympathetic: g. ciliare, g. submandibulare, g. oticum,
g. sphenopalatinum,

Sympathetic system



Parasympathetic system



Different influence of sympathetic system in GUT

Peripheral components of ANS / 2

Mediators (neurotransmission + neurocrine secretion)

Sympathetic - catecholaminergic system (exceptions - vessels, adrenal medulla, sudomotoric seat glands); modulatory NA+ NPY

Parasympathetic - cholinergic system; modulators ACh + SP

Principles

Sym. + Parasymp. act mostly antagonistically; sometimes synergically or independently

Sympathetic - fright- fight or flight (sympathetic arousal)

alpha reaction: fear, fringht (piloerection, vascular constriction, sphincter constriction, pupillar dilation)

beta reaction: action, activity (cardiac stimulation, muscle vasodilatation, bronchodilatation)

Parasympatikus - relaxation (blood supply, increased peristalsis in GUT, salivation, muscle relaxation)

Cental components of ANS / 1

- **Spinal level :**
intermediolateral collumn (intermotoneurons, interosensoric neurons, inhibitory interneurons, etc.)
- **Brainstem :**
 - bulbar visceral-sensoric relay - *nucl. of tractus solitarii*,
 - pontile visceral-sensoric relay - *nucll. parabrachiales*, *nucl. Kölliker-Fuse*,
 - viscerosensoric nuclei & visceromoric nuclei of cranial nerves (III, VII, IX, X)
 - serotonergic (B-group), kachecholaminergic (A, C - groups)
 - vasopressor, vasodepressor, kardiostimulatory, cardioinhibitory reflex circuits (**rostral medulla**)

Central components of ANS /2

- **Subcortical level :**

Hypothalamic-hypophyseal system - somatotopic representation of various qualities of visceral afferentation and efferentation for different body segments of smooth muscles, glands and inner organs

Laesions:

___ **ventromedial hyp.** (tzv. center of satiety) - *polyphagia, bulimia, obesity, lethargia*

ventrolateralis (tzv. center of hunger) - *cachectia, anorexia,*

rostromedial hyp. - *polydipsia, polyuria,*

rostromedial hyp. - *oligodipsia, hyperglycaemia, glykosuria, hypoinsulinaemia*

laterocaudal/dorsal hyp. (tzv. heat production/ conservation center)
paroxysmal hypothermia, poikilothermia,

rostromedial hyp. (tzv. heat dissipation center) - *central neurogenic hyperthermia, hyperhidrosis*

other: progressive lipodystrophy, hypoalbuminaemia, hemorrhagic erosions or ulcers in esophagus, stomach and duodenum

Disorders of autonomic nervous system

Central components of ANS /3

- **Cortical level:**

- **Viscerosensory :** *medial area of temporal-parietal cortex*

Parasympathetic representation - *insula (area 13), temporoparietal cortex (area 38), subparietal region of g. cinguli (area 23, 31);*

laesion → overactivity of sympathetic sy, mydriasis, tachycardia, bronchodilatation, piloerection, decreased peristalsis in GUT, etc.)

Sympathetic representation - *temporal- basal cortex (area 20, 36, 37, 38), perisplenic area (area 26, 29, 30);*

lézie → prevaha parasympatika (napr. mioza, vazodilatácia, bradykardia, hypotenzia, bronchokonstrikcia, hyperperistaltika čriev, hypersekrécia slín, potu a hlienu a iné).

- **Visceromotorika:** *anterior and basal frontal cortex (area 10, 11, 12, 47), anterior part of insulae (area 14, 15, 16) resp. subfrontal part of g.cinguli (area 24, 25, 32, 33).*

laesion → smooth muscle disorders

Types of dysautonomias

- According to etiopathogenesis:

a) Primary vegetative disorders - without proven organic damage (e.g. primary autonomic failure)

b) Secondary vegetative disorders - consequence of acute and chronic diseases (e.g. napr. diabetic and uremic neuropathy)

- According to location and/or mechanisms:

a) Topic syndromes - on particular levels of central and peripheral autonomic regulation,

b) Reflex syndromes - involvement of vegetative endings from various peripheral organic lesions,

c) Global syndromes - systemic disorders involving either sympathetic or parasympathetic system or both

Various signs of dysautonomia / 1

1. Psychological disorders

phobias, anxiety, aggression, tiredness, decrease of intellectual output, sleep disorders, and others

2. Circulatory disorders

- **Microcirculation:** sudden redness or paleness of skin (flush), cyanosis or edematous manifestation without organic reasons, dermatographism, etc.

- **Systemic circulation:** orthostatic hypotension, collapse, hypertension, paroxysmal cardiac dysrhythmia (tachycardias, bradycardias, extrasystoles), angina pain, neurocirculatory asthenia,

- **Cerebrálna cirkulácia:** vasomotor headache, vertigo, fainting, syncope, zvýšená meteorosenzitivita, migraine atď.

Various signs of dysautonomia / 3

7. Secretory disorders

hyperhidrosis or anhidrosis, hyposalivation or hypersalivation, hypolacrimation or hyperlacrimation, lactation disorders, change in composition of sweat, saliva or mucus in respiratory airways, etc.

8. Disorders of regulation of body temperature

hyperthermia, feeling of cold, subfebrility, poikilothermia etc.,

9. Poruchy metabolizmu

kachectia, anorexia nervosa, adiposity independent of food intake, disorders of metabolism of fat, acute hyperglycaemic or hypoglycaemic attacks, etc.

Various signs of dysautonomia / 2

3. Respiratory disturbances

respiratory dysrhythmias, secondary hyperpnoea, tachypnoea, laryngeal stridor, dysphonia, bronchospasms, neurogenic asthma,

4. Gastrointestinal disorders

dysphagia, hypomotility or hypermotility of bowels, spasms in stomach, neurogenic (psychogenic) diarrhoea, constipation, meteorism, insufficiency or spasms of sphincters (cardia, pylorus, rectum), abdominal cramps w/o organic reasons

5. Urinary disorders

polyuria, anuria, nyktúria, urinary urgency, incontinency, painfull spasms of urinary sphincters

6. Genical disorders

sy. of erectile dysfunction, penile intumescence, dyspareunia, menstruation disorders

Various signs of dysautonomia / 4

10. Endocrine disorders

sexual hormonal dysfunction, thyreoidal & parathyreoidal disorders, dysbalance in adrenal functions, pheochromocytoma, etc.

11. Trophic disorders

atrophy and keratosis of skin, skin pigmentation disorders, loss of hairness, demineralisation of skeleton, muscle, ligamental and fascial calcinosis, bone necrosis, neurogenic allergy (urticaria, Quincke oedema), atrophy of testes, etc.

12. Disorders of blood and immune cells

disorders of trombocytic aggregation, reactive anaemias, T-lymphocyte dysfunctions, macrophage dysfunctions, etc.

Common autonomic disorders

Multiple systemic atrophy (MSA)/ Shy-Drager sy. (familial orthostatic hypotension)

- **Characteristics:** fall of BP $> 20/10$ mmHg during sudden change of body position from sitting or lying position to standing position, transient rise of HR > 30 /min
- **Mechanism:** progressive disappearance of autonomic functions
- **Signs:**
 - attacks of severe postural hypotension - syncopas,
 - parkinsonism, cerebellar ataxia, central sleep apnoea
 - other autonomic dysfunction (anhidrosis, erectile dysfunction, dysuria, disorders of peristalsis)
- **Etiology:** progressive degeneration and atrophy of cortex and other regions in CNS

Sy. of orthostatic (postural) hypotension

- **Characteristics:** fall of BP $> 20/10$ mmHg during sudden change of body position from sitting or lying position to standing position, transient rise of HR > 30 /min
- **Occurrence:** both in women and men 20 - 50 y.
- **Mechanism:** failure of sympatheticotonic postural baroreflex
- **Signs:** sudden fainting or fall, paleness in face, vertigo and titubations - loss of balance, headache, tiredness
- **Etiology: hereditary- congenital;**
 - metabolic defects in catecholamine synthesis,
 - vascular alpha-adrenergic receptor hyposensitivity

Sy. of orthostatic intolerance/ Sy. of postural tachycardia (POTS)

- **Characteristics:** change of BP $< 20/10$ mmHg during posturation, rise of HR > 30 /min and plasmatic level of NE 600 pg/ml during longer standing
- **Occurrence:** young women < 35 r. (F: M = 4:1)
- **Mechanism:** permanently increased basal sympatheticotonia
- **Signs:** palpitations, weakness, fainting, redness and swelling of lower extremities, scotomas, tremor, headache, tiredness during long duration standing
- **Etiology: unknown;**
 - (a) preceding viral infection?,
 - (b) cardiac beta-adrenergic receptor hypersensitivity,
 - loss of cardiac baroreflex sensitivity;
 - autoimmune: Ab against alpha-3 nicotine receptor in ganglia

Sy. of ortostatic intolerance/ Sy. of postural tachycardia (POTS)

Hyperadrenergic orthostatic hypotension	Partial dysautonomia
Orthostatic tachycardia syndrome	Sympathotonic orthostatic hypotension
Postural orthostatic tachycardia syndrome (POTS)	Mitral valve prolapse syndrome
Postural tachycardia syndrome	Soldier's heart
Hyperadrenergic postural hypotension	Vasoregulatory asthenia
Sympathotonic orthostatic hypotension	Neurocirculatory asthenia
Hyperdynamic beta-adrenergic state	Irritable heart
Idiopathic hypovolemia	Orthostatic anemia