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Resource description	This lecture is on syndromes in dentistry.
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SYNDROMES

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26. Reye's Syndrome:
27. Grave's Disease:
28. Von Recklinghausen Syndrome or Neurofibromatosis

Eagle Syndrome

Pierre Robin Syndrome

Plummer Vinson syndrome

1) Gardner's Syndrome:- Congenital Diseases

- Multiple osteomas (particularly of jaws and facial bones)
- Multiple **poylps** of large intestine
- Epidermoid Cysts
- Fibromas of the skin

2) Down Syndrome (genetic) also called TRISOMY 21

- Facial Appearance Brachycepaly
- Mid face retrusion
- Small nose with flattened nasal bridge
- Mongoloid Slant

- Macroglossia
- Delayed eruption of teeth
- Major relevant associations are heart defects, atlanto axial subluxation, and anemia and increased risk of leukemia.

3) Ramsay Hunt Syndrome:-

It is thought to be due **herpes zoster** of the geniculate ganglion.

Characteristics:

- Lower motor neuron facial palsy on the same side in the pharynx involves external auditory canal and on the face.

4) Sjogren's Syndrome

- Autoimmune chronic inflammatory disease involving salivary and lacrimal glands. Sjogren syndrome can have widespread manisfestation and is classified into:-

A) Primary Sjogren's syndrome:-

- Also called **SICCA SYNDROME**
- Xerostomia (dry mouth)
- Kerato conjunctivitis sicca i.e. (dry eyes)

5) Gorlin Goltz Syndrome:- Multiple basal cell naevi syndrome characterized by:

- Multiple basal cell carcinoma
- Multiple jaw cysts (**odontogenic keratocyts**)
- Vertebral and rib abnomalities (BIFID RIBS)
- Calcification of falx cerebra
- Frontal bossing
- Mandibular prognathism
- Hypertelorism
- Hydrocephaly
- Eye and endocrine abnormalities.

6) Secondary SJOGREN Syndrome

- In addition to xerostomia (**dry mouth**) and xerophthalmia (**dry eyes**), this has association of AUTO IMMUNE **CONNECTIVE** tissue diseases.
- Autoimmune connective tissue diseases in secondary Sjogren's syndrome.

-Rheumatoid diseases (**arthritis**)

-Systemic lupus erythematosus

-Progressive systemic sclerosis

-Primary biliary cirrhosis

-Renal tubular acidois

-Mixed connective tissue disorder.



Cleidocranial Dysostosis

Cleidocranial Dysostosis:

-Autosomal dominant inherited condition
- Following are the characteristic features:

- Hypoplasia or aplasia of clavicles
- **Delayed ossification of cranial fontanelles**
- A large, short skull

- Shortness of stature
- Frontal and parietal bossing
- Failure to pneumatize the air sinuses
- **Highly arched palate** and clefting
- Mid face Hypoplasia
- Failure of tooth eruption with multiple supernumerary teeth
- Teeth has abnormalities such as gemmination, hypoplasia of secondary cementum may occur.

8) Ectodermal Dysplasia:-

- Usually sex linked reccesive trait.
- Hypodontia more often most of the decidous teeth form but there are few or no permanent teeth. Teeth are peg shaped or conical.
- **Hypotrichosis-** (scanty hair)
- Inability to sweat skin is smooth, shiny and dry due to absence of sweat glands.

9) Grinspan&#xfffd;s Syndrome

Characterized by presence of the triad:

- **Hypertension**
- **Diabetes mellitus**
- **Oral lichen planus**
- Oral lichen planus is thought to be a result of drug used for hypertension and diabetes mellitus but this is not confirmed.

10) Cleft Lip and Palate:

- Clefting of the lip and primary palate follows failure of fusion of the medial nasal, lateral nasal and maxillary processes at around **6th week** of intrauterine life.

- Cleft of secondary palate follows failure of fusion of palatal shelves from 8-10 weeks.

CLASSIFICATION ✓ several classifications exist:-

I. Primary(Lip and alveolus to the incisive foramen) or
Secondary (Hard palate from incisive foramen back and soft palate)

II. Unilateral or Bilateral

III. Complete or Incomplete

INCIDENCE OF DENTOFACIAL CLEFTING

BIFID Uvular	1:100
ISOLATED cleft lip	1.0000 (either unilateral /bilateral)
ISOLATED cleft palate	1:2200
COMPLETE cleft	1:1800

Cleft lip and palate more frequently affects males and more common on left than on **right side.**

AGE:	TREATMENT
Birth	Typical sequence of treatment for patients with cleft lip and palate. Initial assessment, preclinical orthodontics is no longer carried out.
3 months	Primary lip repair surgery; Millard and Delaire are two commonly used, surgical techniques.
9-18 months	Surgical repair of palate, which is good for speech development but is associated with impaired growth of the maxilla; von langenbeck and Delaire are two commonly used surgical techniques.
2 years	Speech assessment

3-5 years	Lip remission surgery
8-9 years	Pre bone graft orthodontic treatment; speech therapy.
10 years	Alveolar augmentation with cancellous bone from ideal crest, which allows maxillary canines to erupt and provides support for a alveolar base
12-14 years	Definitive orthodontics
16 years	Nose revision surgery
17-20 years	Advanced conservation treatment, orthographic surgery to correct, hypoplastic maxilla

11) Frey Syndrome:

- It follows trauma to skin overlying a salivary gland (parotid gland) and is thought to be due to post-traumatic crossover of sympathetic and parasympathetic innervation to the gland and skin respectively.

Signs and symptoms:

- Erythema (redness/flushing)
- **Sweating** in the cutaneous distribution of the auriculotemporal nerve, usually in response to gustatory stimuli.

12) BEHCET SYNDROME DEFINITION:

Signs and Symptoms

- **Oral ulceration**
- **Genital ulceration**
- **Uveitis**

Commonly affects young adults, especially **male**.

13) MELKERSON ROSENTHAL SYNDROME

Consists of:

- Facial paralysis
- Facial oedema
- Fissured tongue

14) OROFACIAL GRANULOMATOSIS

Signs and Symptoms:

- Persistent or recurrent enlargement of the lips, causing them to protrude.
- Midline fissuring of lip (**Median Cheilitis**)
- Sores of corner of mouth (**Angular Cheilitis**)
- Oral ulcers
- Marginal Gingivitis
- Fissured tongue
- Enlargement of peri-oral and peri-orbital soft tissues
- Cervical lymphadenopathy
- Facial palsy

ALBRIGHT SYNDROME

- Consists of polyostotic fibrous dysplasia
- Patchy skin pigmentation (**caf-au-lait spots**)

- Endocrine abnormality

CHEDIAK- HIGASHI SYNDROME:

- Defective neutrophil function
- Abnormal skin pigmentation
- Increased susceptibility to infection (severe gingivitis, periodontitis, aphthae in young children)

CROUZON SYNDROME:

- Autosomal dominant
- Premature fusion of cranial suture, mid face hypoplasia
- Proptosis of the globe of the eye.
- Radiologically **beaten copper skull**
- Increased intracranial pressure
- Risk of blindness.

EHLERS DANLOS SYNDROME:

- **Hyper flexibility of joints**
- Increased bleeding and bruising
- Hyper extensible skin
- Bleeding is common in type IV
- Early onset periodontal disease in type VIII
- **Pulp stones** may be seen in all types.

PAPILLON- LEFEVRE SYNDROME:

- Palmoplantar hyperkeratosis & juvenile periodontitis which affects both 1 & 2 degree dentition.
- **Aggressive gingivitis & periodontitis**

PATTERSON- BROWN- KELLY SYNDROME(Plummer-vinson syndrome)

- **Dysphagia**
- Microcytic hypochromic **anemia**
- Koilonychia (spoon shaped nails)
- **Angular Collincheilitis**
- Affected group is middle aged women

PEUTZ-JEGHERS SYNDROME:

- Autosomal dominant condition of melanotic pigmentation of skin (especially peri-oral skin), mucosa & **intestinal polyposis.**
- Ovarian tumors are sometimes associated with the condition (10% woman with Peutz-Jeghers)

TREACHER-COLLIN SYNDROME:

- Defects in structure derived from the first brachial arch
- Autosomal dominant
- Down-ward-sloping (antimongoloid slant) palpebral fissure
- Hypoplastic malar complexes
- Mandibular retrognathia with a high genial angle
- Deformed pinnas
- Hypoplastic air sinuses

- Colobomas in outer third of eye
- Middle & inner ear hypoplasia (deafness)
- Cleft palate
- Unusual tongue like projection of hair pointing towards the cheek
- Funny looking kids

Heerfordt syndrome: (Uveoparotid fever)

Is sarcoidosis associated with lacrimal, salivary (parotid swelling), uvetitis and fever. Sometimes it is associated with neuropathies like facial palsy.

Steven- Johnson Syndrome:

A severe version of Erythema multiforme.
A mucocutaneous condition that is probably autoimmune in nature and precipitated particularly by drugs.

Classical signs are Target lesions and Concentric red rings affecting hands and feet.
It is associated with fever and mucosal involvement.
Viral infections, like herpes simplex are the second commonest cause.

Horner Syndrome:

Consists of a constricted pupil (Miosis), drooping eyelid (ptosis) , unilateral loss of sweating (anhydrosis) on the face, occasionally sunken eyes (enophthalmos)

Reye's Syndrome:

It is occur due to aspirin in children under 16 years.

It is fatty degeneration of liver, can be lethal.

Grave's Disease:

It is an auto immune hyperthyroidism
autoantibody- TSH
causes eye signs like exophthalmos, lid lag and lid retraction

Von Recklinghauson Syndrome or Neurofibromatosis

It is an Autosomal dominant disease
It is central nervous syndrome
An autosomal dominant diseases

Characterised by Multiple neurofibromas with skin pigmentation, skeletal abnormalities

Eagle syndrome

- Dysphagia
-Pain on turning head associated with an **elongated styloid process.**

Pierre-robin-syndrome

-Micrognathia

-Cleft Palate

-Glossoptosis

-Upper respiratory obstruction

Hutchinson's triad is named after Sir Jonathan Hutchinson (1828–1913). It is a common pattern of presentation for congenital syphilis, and consists of three phenomena: interstitial keratitis, Hutchinson incisors, and eighth nerve deafness.

PRINCIPAL SOURCES AND FURTHER READING:

-R.J GORLIN 1990 SYNDROMES OF HEAD AND NECK 3RD EDITION
-DOWNS SYNDROME ODELL:CLINICAL PROBLEM SOLVING
-OXFORD HANDBOOK OF CLINICAL DENTISTRY 5TH EDITION

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