the Child Dental Patient with

Fetal Alcoholic Syndrome

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European Reference Network
for rare or low prevalence complex diseases
Network
Craniofacial anomalies and ear, nose and throat disorders (ERN CRANIO)
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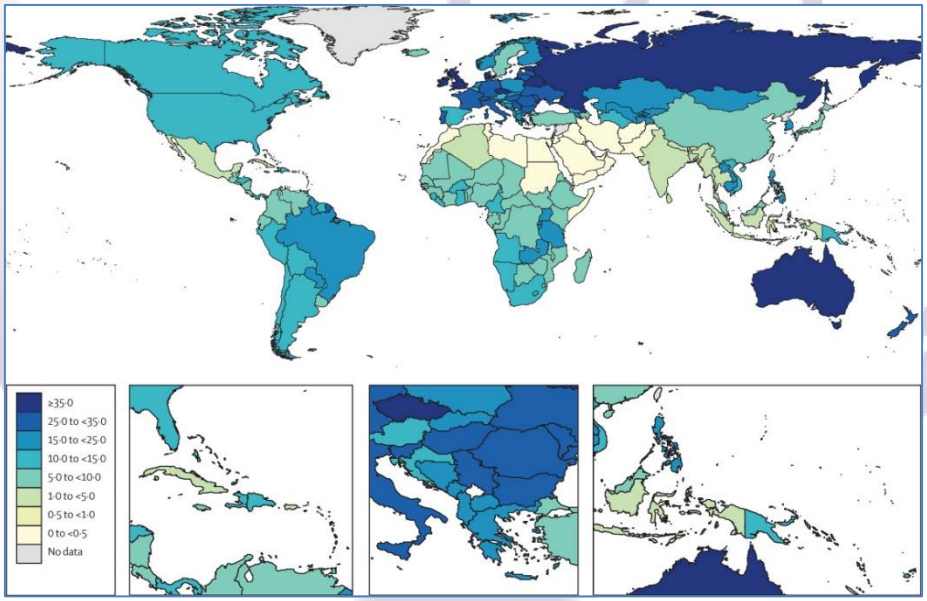
Definition

- Alcohol is a well-established teratogen that can cause variable physical and behavioral effects on the fetus, named as fetal alcohol spectrum disorders (FASD).
- The most severe condition in this spectrum of diseases is known as fetal alcohol syndrome (FAS).

- FAS is a common cause of mental retardation.
- Infants with FAS have prenatal and postnatal growth retardation in weight, height and often head circumference, altered morphogenesis, especially dysmorphic facial features, and central nervous system involvement.

Epidemiology FAS/FASD

- The diagnosis is subdivided into subgroups according to the severity of the symptoms:
  - Estimates of FAS incidence vary considerably, reported as 0.5 to 3/1,000 live births.
  - Fetal alcohol spectrum disorders (FASD) are often undiagnosed (global prevalence = 7,7 per 1000).
- High FASD prevalence reflects the high prevalence of alcohol use and prenatal alcohol exposure: approximately 25% of 18–34 year old men and women binge drink (*defined by as five of more drinks per occasion for men and four or more for women*), and 7,3% of pregnancies are alcohol-exposed.
- Binge drinking by women of childbearing age remains a problem globally and is increasing in some countries.



Popova et al, 2017

Pathogenesis

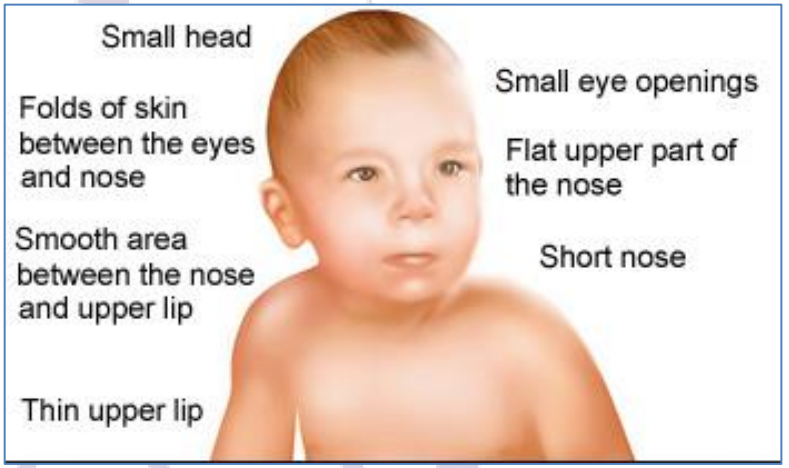
- Alcohol has been a well-established teratogen for many years.
- Both animal and clinical studies have shown that ethanol (EtOH) diffuses through the placenta and distributes rapidly into the fetal compartment where EtOH also has a slower elimination rate, accumulating in the amniotic fluid.
- This reservoir causes greater fetal exposure to EtOH and is compounded by fetal swallowing, caused by the fetal kidneys excreting xenobiotics into the amniotic fluid, which the fetus then swallows.
- Alcohol therefore has a prolonged effect on the fetus due to amniotic accumulation, reduced concentrations of fetal metabolic enzymes, and reduced elimination.

Factors increasing risk of FAS/FASD

- The risk of developing FAS is related to timing and amount of alcohol consumption (dose-dependent):
  - greater numbers of drinking days per week and sustained drinking throughout all trimesters increase the risk
  - the first trimester is the most vulnerable time period with a 12-times increased risk .
- Various maternal characteristics also influence FAS risk:
  - smaller body profile (height, weight), body mass index
  - poor nutritional status
  - low socioeconomic status and smoking.
- Other risk factors include genetic influences and paternal chronic alcohol use.

Clinical features

- **Craniofacial dysmorphology**:
  - short palpebral fissures
  - smooth philtrum
  - thin upper lip vermilion
  - these dysmorphic features are clinically detectable in a minority of cases
  - diagnosis of smooth philtrum and thin upper lip can be performed using the Lip-philtrum guide by Astley and Clarren, which consists of photographs to be visually compared to the patient during diagnosis.
- **Growth deficiency**: Height and/or weight ≤10th centile based on racially/ethnically normed charts
- **Cardiac anomalies**: aberrant great vessels, atrial septal defects, ventricular septal defects
- Delayed in walking
- Gross and fine motor deficits (balance and incoordination)



These motor deficits limit children in performing basic motor skills in everyday life

Clinical features

- **Sensory and neuropsychological abnormalities:**
  - impaired odor identification
  - microphthalmia with associated reduction in palpebral fissure length, coloboma, optic nerve hypoplasia, retinal dysplasia, retinal vascular tortuosity, convergent strabismus, and low visual acuity
  - various forms of hearing loss
  - atypical auditory processing.

These impairments may have greater impact on speech, language, reading, and writing development

Cognitive / behavioural impairments

- **Global cognitive impairment or impaired performance.**
- **Summary of psychological deficits in FAS** (*Jacobson and Jacobson, 2002*)
  - hyperactivity
  - attention deficits—sustained and focused attention
  - planning difficulties
  - learning/memory problems
  - poor consolidation of new memories
  - lower IQ—arithmetic, receptive language, and verbal processing problems
  - social difficulties
  - oppositional defiant disorder.
- **Common secondary disabilities in FAS** (*Mukherjee et al., 2006*)
  - psychiatric problems
  - disrupted school experience
  - trouble with the law
  - confinement
  - inappropriate sexual behaviors
  - substance use disorders.

Brain abnormalities and co-morbidities

- Brain abnormalities:
  - microcephaly
  - neural tube defects
  - hypoplasia of the corpus callosum and cerebellum
  - vascular anomalies
  - focal gliosis
  - perivascular space dilation
  - pituitary hypoplasia
  - ventriculomegaly…

- Co-morbidities /co-occurring conditions:
  - recurrent non febrile seizures (5-9%)
  - sleep disturbances (may contribute to the neurocognitive and behavioral deficits)

Oral characteristics

- Cleft palates and cleft palate and lips.
- Narrow-arched, high-arched.
- Malocclusions
  - overjets, cross-bites, crowded and misaligned teeth
  - could be in relation with lack of breastfeeding, mouthbreathing…
- Modified DDE index (enamel defects) significantly higher.
- Higher DMFT Index in patients with FAS possibly due to reduced motor skills.

Prevention of FAS/FASD

- The most effective method of FAS/FASD prevention is to stop maternal alcohol consumption during pregnancy.
- Prenatal methods to reverse or prevent alcohol's teratogenicity mechanisms are being explored: antioxidants as food supplements, vitamin C , folic acid, L-glutamine, boric acid, choline…

Management of FAS/FASD

- There are no specific drug treatments for FAS/FASD.
- Multi-faceted interventions generally follow recommendations for other developmental disorders in children:
  - specialized educational intervention
  - use combinations of medications for ADHD, disorders of impulse control, aggression, and mood disorders.

Dental management of FAS/FASD

If cardiac malformation, antibioprophylaxis

- Patients with FAS/FASD have to be included in **oral health prevention programs** at a dental office with:
  - a higher number of appointments
  - frequent professional tooth cleaning
  - more intensive training and instruction concerning oral health and tooth brushing methods
  - prescription of fluoride rinses/gels
  - application of fluroride varnishes on a regular basis.

- **Management of anxiety and of behavioural problems:**
  - behaviour management
  - sedation, premedication

- **Prevention of malocclusions**:
  - Identify sucking habits and mouthbreathing at an early stage.
  - An orthodontist, as well as an otorhinolaryngologist, should be consulted at an early age.

Conclusions

- FAS/FASD = global public health problem that remains under-recognized and under-diagnosed despite its high prevalence and cost to society.
- Such patients may have numerous emotional or physical problems that the dentist should recognize.
- The dentist could also contribute to the diagnosis of FAS/FASD by **identification of the facial phenotype.**