

# Oral Health Fact Sheet for Dental Professionals

## Adults with Down syndrome (Trisomy 21)

*Down syndrome is a chromosome disorder associated with an extra chromosome (Trisomy 21) resulting in intellectual disability and specific physical features. (ICD-9 code 758)*

### Prevalence

- < 1%
- Three types of chromosomal abnormalities can lead to Down syndrome:
  - \* Nondisjunction: 95% (males 59%, females 41%)
  - \* Translocation: 4% (females 74%, males 26%)
  - \* Mosaicism: 1% (may have more subtle features)
- Medical complications seem to be similar in all three groups.

### Manifestations

#### Clinical

- Increased risk of abnormalities in almost every organ system
- Intellectual disability and delayed growth
- High incidence of Alzheimer's disease in individuals over 40 years
- Vision and hearing problems
- Cardiac defects (VSD, ASD, PDA, Tetralogy of Fallot)
- Characteristic physical features: brachycephalic skull, prominent epicanthic skin folds, small low-set ears, reduced muscle tone, pelvic dysplasia, transverse palmar crease, broad hands and feet, short fingers, and lenticular opacities

#### Oral

- Early onset severe periodontal disease (most significant oral health problem)
- Adults with Down syndrome are at an increased risk of caries due to xerostomia and cariogenic food choices. Hypotonia contributes to chewing problems and inefficient natural cleansing action.
- Congenitally missing and malformed teeth are common; malocclusion
- Hypoplasia of mid-facial region
- Hypodontia, microdontia
- Macroglossia, fissured and protruding tongue
- Tongue thrust, bruxism, clenching, mouth breathing

#### Other Potential Disorders/Concerns

- Epilepsy
- Cardiac defects
- Atlantoaxial instability (fragility of cervical vertebrae/spinal cord)
- Compromised immune system
- Sleep apnea
- Increased risk of leukemia
- Hearing loss
- Vision problems
- Hypothyroidism
- Memory loss and dementia
- Dry skin

# Adults with Down syndrome (Trisomy 21) continued

## Management

The list of medications below are intended to serve only as a guide to facilitate the dental professional's understanding of medications that can be used for Down syndrome or conditions associated with Down syndrome. Medical protocols can vary for individuals with Down syndrome from few to multiple medications.

Medications are prescribed based on symptoms:

SYMPTOM	MEDICATION	SIDE EFFECTS/DRUG INTERACTIONS
Seizures	<b>Anticonvulsants</b>	
	<i>Carbamazepine</i> (Tegretol)	Xerostomia, stomatitis, glossitis, dysgeusia, bone marrow suppression. Excessive bleeding may result when combined with aspirin or NSAIDs.
	<i>Valproate</i> (Depakote, Depakene)	Xerostomia, stomatitis, glossitis, dysgeusia, oral petechia. Excessive bleeding may result when combined with aspirin or NSAIDs.
	<i>Phenytoin</i> (Dilantin)	Xerostomia, gingival hyperplasia.
	<i>Gabapentin</i> (Neurontin)	Xerostomia, fever, mood changes, erythema multiforme, kidney failure, thrombocytopenia, viral infections, hyperkinesia, other neurologic symptoms.
	<i>Levetiracetam</i> (Keppra)	Hostility, irritability, mood changes, depression, anorexia, infection, gingivitis.
	<i>Lamotrigine</i> (Lamictal)	Angioedema of mouth, lips, tongue or face; oral lesions, xerostomia, nausea, headache, blurred vision, double vision, Stevens-Johnson syndrome (uncommon, severe).
Muscle Spasticity and Rigidity	<b>Muscle relaxants and antispasmodics</b>	
	<i>Baclofen</i> (Lioresal)	Xerostomia (uncommon), angioedema of mouth, lips, tongue or face.
	<i>Diazepam</i> (Valium)	Drowsiness, dystonia, double vision, xerostomia or hypersalivation, seizures, CNS and respiratory depression, paradoxical CNS stimulation, tiredness, syncope, fatigue, ataxia, depression, headache, nausea. Alcohol, and drugs that cause sedation, may increase the sedative effect of diazepam. Use with caution for persons with sleep apnea.
	<i>Dantrolene sodium</i> (Dantrium)	Drowsiness (alcohol can increase this effect), weakness, dizziness, tachycardia (increased heart rate), abnormal blood pressure, diarrhea, constipation, liver failure. Use caution in combining with drugs that cause CNS depression.

# Adults with Down syndrome (Trisomy 21) continued

SYMPTOM	MEDICATION	SIDE EFFECTS/DRUG INTERACTIONS
Muscle Spasticity and Rigidity <i>continued</i>	<i>Tizanidine</i> (Zanaflex, Sirdalud)	Drowsiness (alcohol can increase this effect), xerostomia, dizziness, hypotension, weakness, somnolence. Do not prescribe with ciprofloxacin or fluvoxamine. Fluoroquinolone antibiotics such as floxacin and norfloxacin interfere with tizanidine metabolism. Use caution in combining with drugs that cause CNS depression.
Hypertension	<i>Verapamil</i> (Calan) <i>Diltiazem</i> (Cardizem, Dilacor) <i>Nifedipine</i> (Procardia) <i>Nicardipine</i> (Cardene) <i>Isradipine</i> (DynaCirc) <i>Amlodipine</i> (Norvasc)	Low blood pressure, gingival enlargement (except isradipine). Verapamil and diltiazem taken with macrolide antibiotics (azithromycin, erythromycin, clarithromycin) may produce prolonged QT intervals and cardiac toxicity. Verapamil and diltiazem inhibit metabolism of benzodiazepines.

## Behavioral: Many individuals with Down syndrome can successfully be treated in the dental office.

### Guidance:

- Plan a pre-appointment (in person/phone) to discuss patient special needs prior to the first visit. Determine if there is a caregiver. Discuss patient's needs with the caregiver.
- Schedule appointments early in the morning or best time of day for patient.
- Talk with the parent or caregiver to determine the patient's level of intellectual and functional abilities and explain each procedure at a level the patient can understand. Most patients with Down syndrome understand more than they can verbalize.
- Use short, clear instructions and speak directly to the patient. Repeat, using the same terms, to compensate for short-term memory difficulties.
- Minimize distractions, such as sights and sounds, which may make it difficult for the patient to cooperate.
- Start the oral examination slowly, using only fingers at first. If this is successful, begin using dental instruments.
- Use the Tell-Show-Do approach when introducing new instruments or procedures. As appropriate, provide verbal and/or tactile reassurances.
- Reward cooperative behavior with positive verbal reinforcement. Consider positive reinforcement at the end of the appointment (e.g., new toothbrush, tube of toothpaste).
- Develop trust and consistency between the dental staff and the patient. Use the same staff, dental operator, and appointment time each visit if appropriate.

## Dental Treatment and Prevention

- Determine who can give informed consent. Be sure they are present at the appointment.
- Consider patient's cardiac status and need for premedication-medical consult may be indicated. A majority of patients with Down syndrome develop mitral valve prolapse by adulthood.
- Ask caregiver for medication updates at each appointment. Medication changes can affect the appropriate care of the patient from a medical and/or appointment management standpoint.
- Monitor periodontal disease closely. A compromised immune system contributes to the high incidence of periodontal disease in the population. The existence of teeth with short conical roots leads to early loss of teeth from periodontal disease. Early treatment is essential. Consider specialty referral if indicated.

## Adults with Down syndrome (Trisomy 21) continued

- Partial and complete dentures may not be well tolerated in some patients due to macroglossia and protruding tongue.
- Powered toothbrushes may be too stimulating for some adults and should be recommended only after determining if the adult will tolerate one.
- Consider daily use of Chlorhexidine or other antimicrobial agents. Evaluate patient – those with swallowing difficulties or inability to expectorate may benefit from brushing teeth and gums with Chlorhexidine or use Chlorhexidine spray application rather than a rinse.
- Evaluate malocclusions. Some patients are good candidates for full orthodontic treatment. Maintain primary teeth (if present) as long as possible and consider space maintenance and orthodontic consultation for missing teeth.
- If tolerated, consider recommending a power toothbrush.

### Considerations due to associated medical conditions:

- Increased gag reflex during oral examination.
- Chronic respiratory infections and open mouth posture → frequent mouth breathing → xerostomia.
- Seizure management during treatment: **Remove** all dental instruments from the mouth. **Clear** the area around the dental chair. **Stay** with the patient and turn patient to one side. **Monitor** airway to reduce risk of aspiration. **Note time** seizure begins: if seizure continues >3 mins., call **EMS** – Danger of Status Epilepticus (potentially life threatening).
- Carefully move patients with atlantoaxial instability into the dental chair, giving special attention to the spine and neck. Use pillows to stabilize the patient and increase comfort, as directed by the caregiver.

### As needed for patients with xerostomia:

- Educate on proper oral hygiene (brushing, flossing) and nutrition.
- Recommend brushing teeth with a fluoride containing dentifrice before bedtime. After brushing, apply neutral 1.1% fluoride gel (e.g., Prevident 5000 gel) in trays or by brush for 2 minutes. Instruct patient to spit out excess gel and NOT to rinse with water, eat or drink before going to bed.
- Recommend xylitol mints, lozenges, and/or gum to stimulate saliva production and caries resistance.

Look for signs of physical abuse during the examination. Note findings in chart and report any suspected abuse to 1-866-ENDHARM ([www.dshs.wa.gov/endharm.shtml](http://www.dshs.wa.gov/endharm.shtml)) as required by law. Abuse is more common in people with developmental disabilities and often manifests in oral trauma.

**Additional information:** [Special Needs Fact Sheets for Providers and Caregivers](#)

# Adults with Down syndrome (Trisomy 21) continued

Below are references and resources which, although some are labeled for children, are very helpful for reviewing implications in adults. Down syndrome manifests at birth with lifelong implications impacting the delivery of dental care.

## References

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- Seagriff-Curtin, P., Pugliese, S., Romer, M. Dental considerations for individuals with Down syndrome. (2006) *NY State Dent J* 72(2):33–5.
- Goldstein, G. Task force on special dentistry makes meaningful improvement. (2006) *NY State Dent J* 72(2):24–6.
- [NIH Institute for Down syndrome](#)

## Additional Resources

- [NIH Institute for Down syndrome](#)
- [ASTDD-Special Needs](#)
- [Block Oral Disease, MA](#)
- Free of charge CDE courses: [NIDCR CDE](#) (2 CDE hours)



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