

Attention and Behavior Problems in Down Syndrome: *Could It Be Sleep Apnea?*

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Introduction

- Historical/Philosophical Perspective
- Intellectual Development in DS
- Review of Sleep Disturbances
- What is Sleep Apnea/Why Is it Important?
- What Causes Sleep Apnea in DS?
- How Is OSA Diagnosed?
- How Can OSA Be Treated?
- Questions and (Hopefully) Answers

If you treat an individual as she is,
she will stay as she is.

But if you treat him as if he were
what he ought to be and could be, he
will become what he ought to be and
could be.

Goethe

Wolraich, M. (ed), Disorders of Development and Learning, Mosby, 1998

Culture Matters—From GB 1998

“There is no effective treatment for Down syndrome aside from the management of infections and the hearing deficit and surgical treatment of correctable associated malformations. An adapted educational programme should be devised for each individual patient. Many children can be maintained within the family but the overall outlook for full autonomy is poor.”

Aicardi, J. et al (Ed) Diseases of the Nervous System in Childhood, Cambridge, 1998

Culture Matters—From the US 1996

Anticipatory health management (Screening):

- Cardiac
- Visual
- Hearing
- Thyroid
- C-spine

Culture Matters—From the US 2010

Anticipatory health management (Screening):

- Cardiac
- Visual
- Hearing
- Thyroid
- C-spine
- ADHD/Behavioral/Emotional/Intellectual
- Alzheimers
- Sleep Disturbances

Common Sleep Disturbances

- Bedtime Resistance
- Sleep Onset Delay—Insomnia
- Sleep Duration
- Sleep Anxiety
- Night Wakenings
- Parasomnias (Sleep walking, nightmares)
- Sleep disordered Breathing e.g. Sleep Apnea
- Daytime Sleepiness

Sleep Disturbance	Children w/DS ages 4-12 years	TD Children ages 4-12 years
Bedtime Resistance	9.75*	7.06
Sleep Onset Delay	1.67*	1.25
Sleep Duration	3.88	3.41
Sleep anxiety	5.92*	4.89
Night awakenings	5.75*	3.51
Parasomnias	11.46*	8.11
SDB	5.75*	3.24
Daytime Sleepiness	15.2*	9.64

Factors Affecting Cognitive & Behavioral Development in DS

- Hypothyroidism
- Hearing Loss (progressive)
- Unrepaired or Evolving Cardiac Disease
- Visual Acuity
- Social/Educational Environment (Low expectations)
- Seizures
- Depression
- Sleep Disturbances/Sleep Disordered Breathing

These are the same for the general population and must be identified and treated in children w/DS with same vigilance for optimal outcomes.

Profound Effects of Sleep Disturbances

- Daytime mood disorders
- Behavioral problems
- Cognition Deficits
- General Performance School/Work
- Family Relationships
 - Maternal stress, depression
 - Poor marital relations
 - Child Abuse
- Physical/Physiological:
 - Pulmonary Hypertension
 - Accelerated Atherosclerotic Heart Disease—
Stroke and Heart Attacks

What Is Sleep Disordered Breathing a/k/a Sleep Apnea?

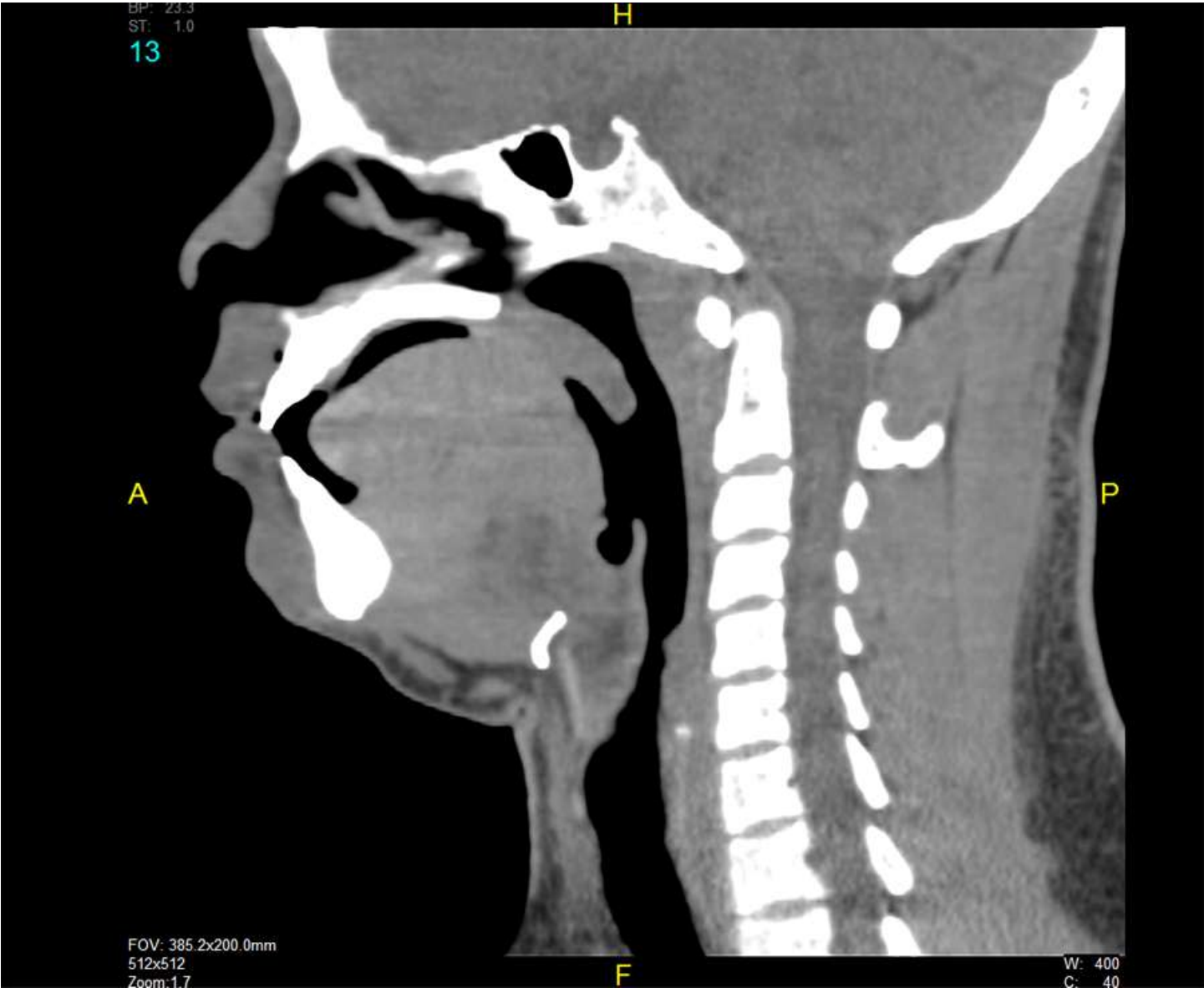
- Obstructive apnea occurs when there is total cessation of airflow for 2 or more respiratory cycles
- Partial obstructions/hypopneas <50% airflow with a drop in oxygen or arousal following event.
- Respiratory Disturbance Index #
apneas+hypopneas/hour
- Hypercarbia—rise in carbon dioxide (not good)
- Hypoxemia—sustained fall in oxygen <92%
- Arousal Index—arousals/hour sleep

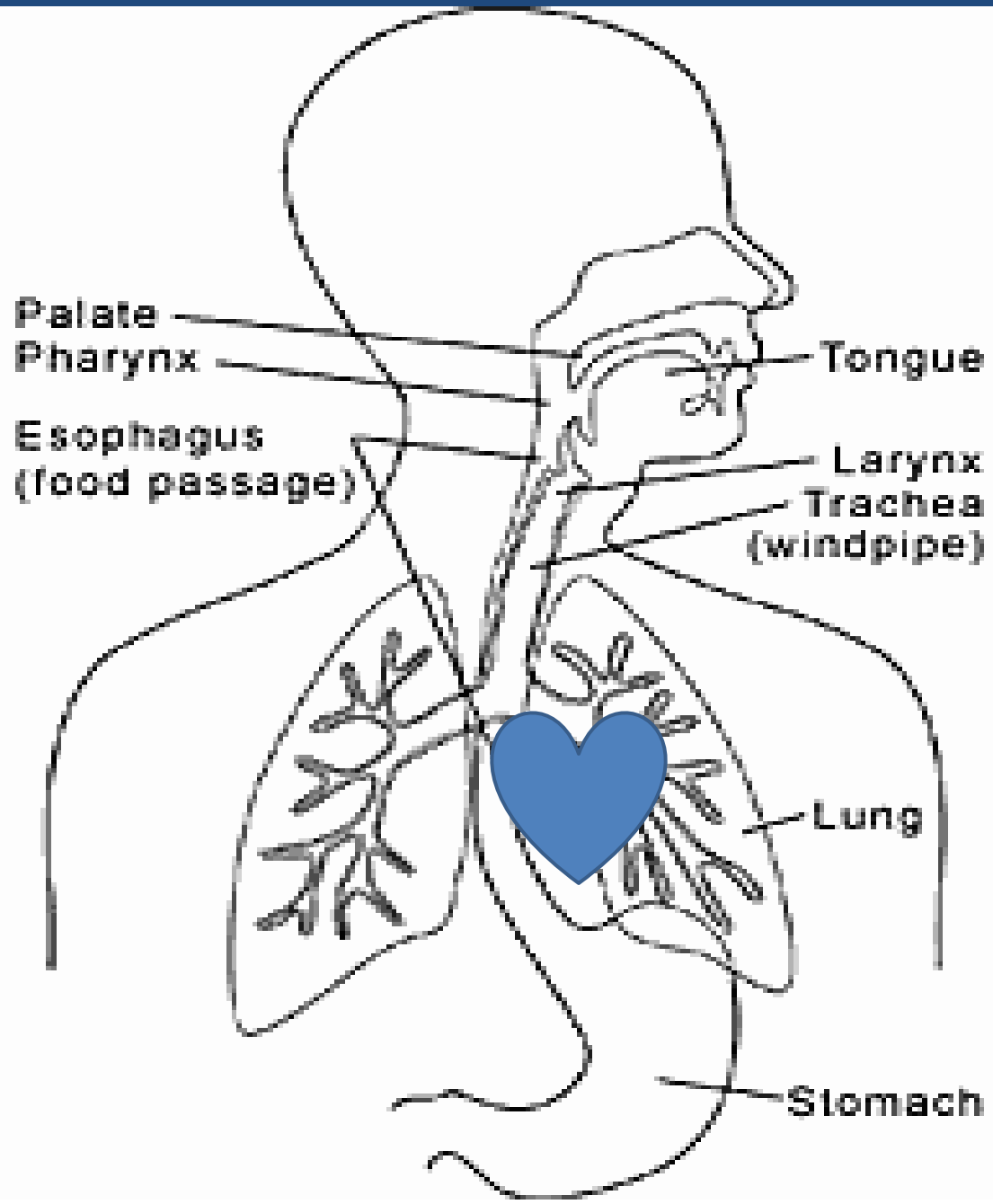
SDB/OSA in DS

- 60-100% DS have SDB
- Sleep disordered breathing and obstructive sleep apnea used interchangeably
- Manifestations:
 - Range from "not-so-benign" snoring to life threatening cardiac failure
 - Partial and total obstructions change the hemodynamic pressures in the thorax (chest)
 - Disrupt normal sleep patterns

Anatomic and Physiologic Factors in DS

- Small, flat mid-face and nasopharynx
- Nasal cavity small, often inflamed/crusted
- Small oral cavity with relatively larger tonsils/adenoids and retro-displaced tongue
- Relative tongue enlargement
- Muscle hypotonia → airway collapse
- Extra-esophageal Reflux
- Obesity





Symptoms of OSA

- Snoring
- Irregular breathing
- Noisy breathing
- Restless sleep/unusual positions
- New onset bedwetting
- Behavior changes
- Daytime sleepiness
- Daytime hyperactivity
- Failure to thrive

Physical Signs of OSA in Children

- Mouth breathing
- Large tonsils/adenoids
- Malocclusion
- Muscular hypotonia
- Thick neck
- Craniofacial—large tongue, small jaw/midface
- Obesity
- Underweight

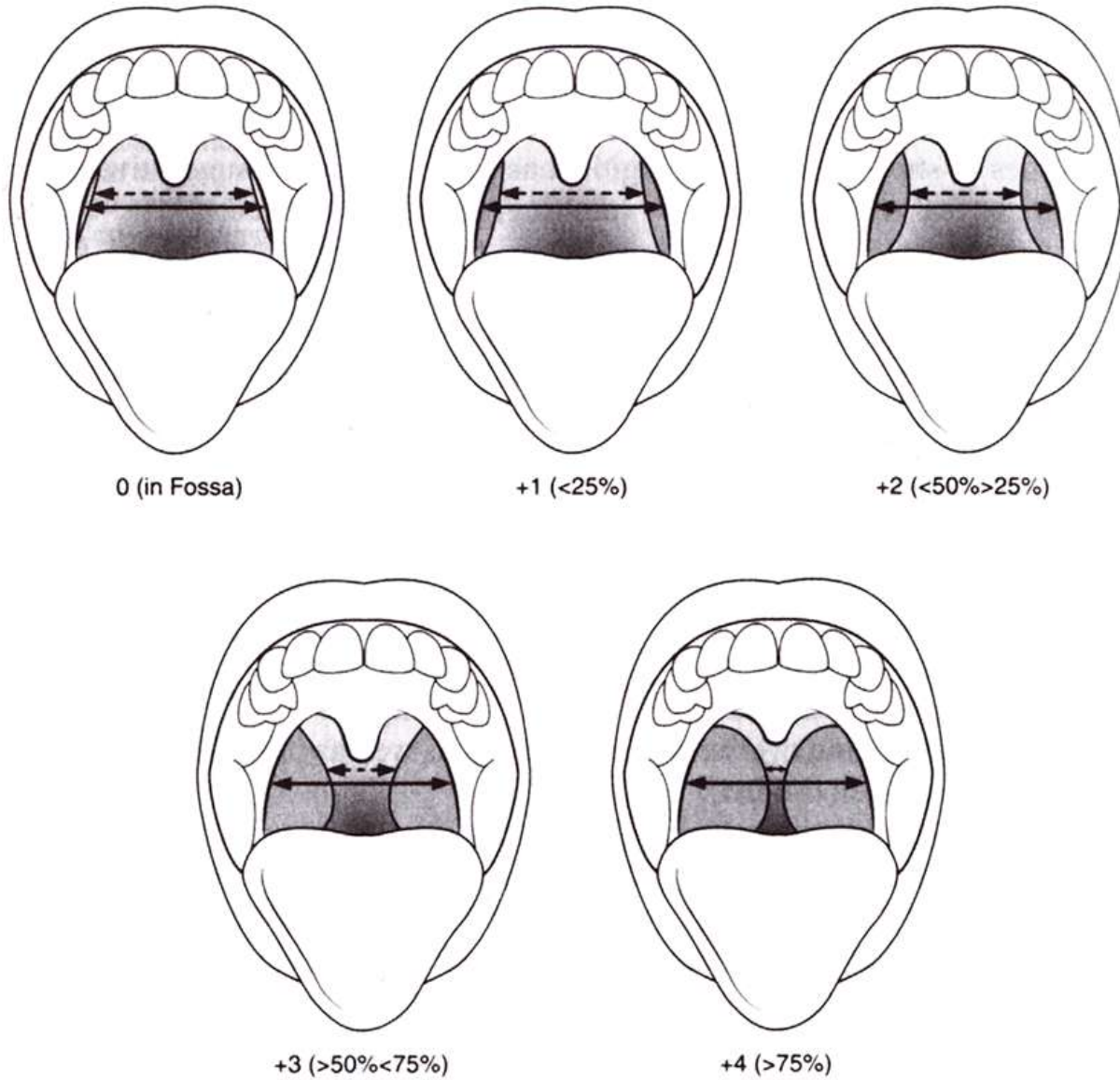
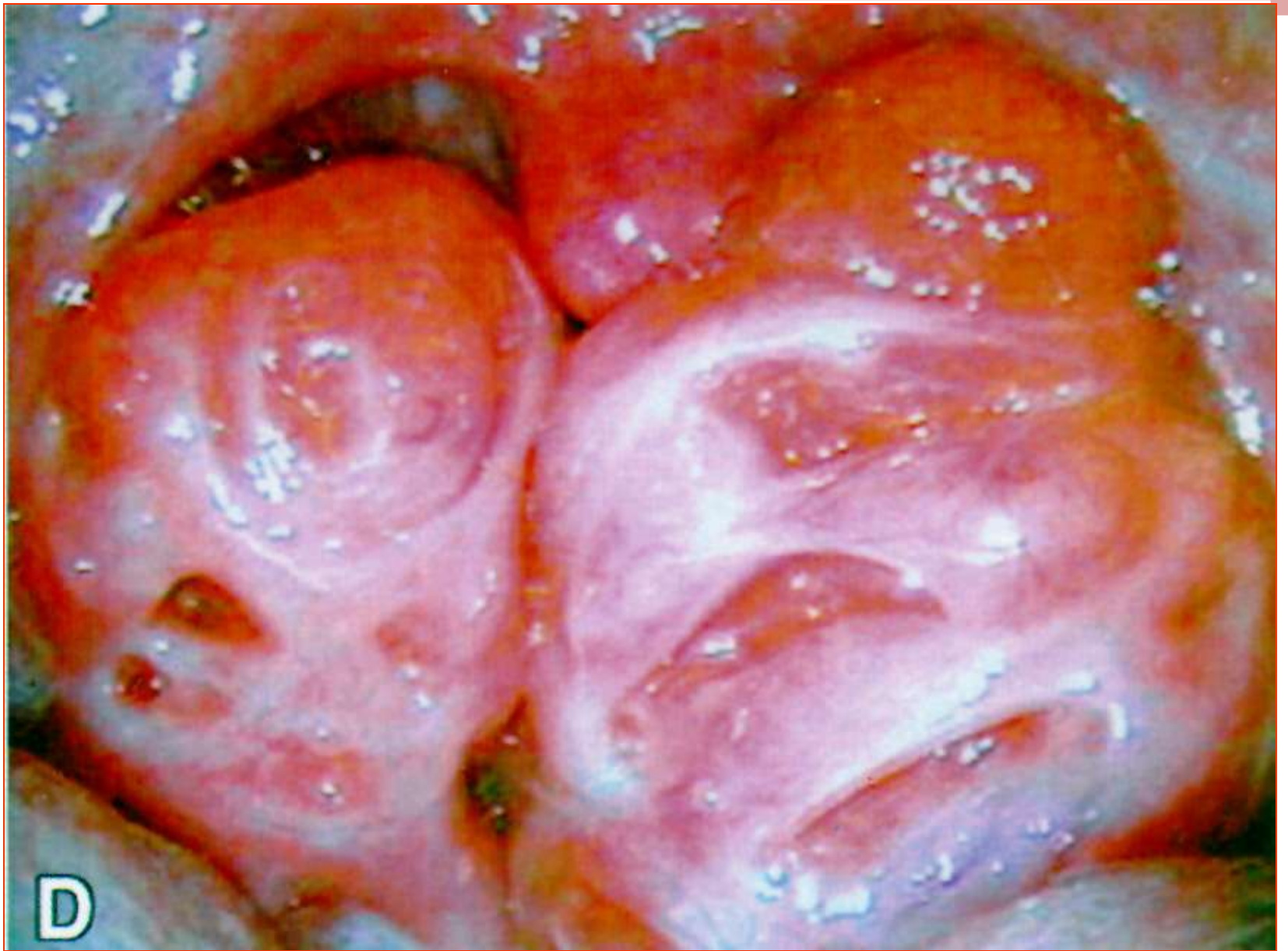


FIG 14.1. Grading tonsillar size.





Laboratory Evaluation

Polysomnogram: "Sleep Study"

Overnight test which measures:

1. Stages of sleep
2. Breathing patterns
3. Blood oxygenation
4. Blood CO₂ levels
5. EEG
6. EKG
7. Limb movement
8. Eye movements

- Obstructive apnea: cessation of airflow for > 8 seconds with persistent inspiratory effort
- Hypopnea: discrete episode of 50% reduction of baseline in airflow with inspiratory flow limitation lasting more than 2 breaths
- OSA in children is defined as an apnea hypopnea index (AHI) >1
- The severity of OSA:
 - mild AHI 1-4,
 - moderate AHI 5-10
 - severe AHI >10



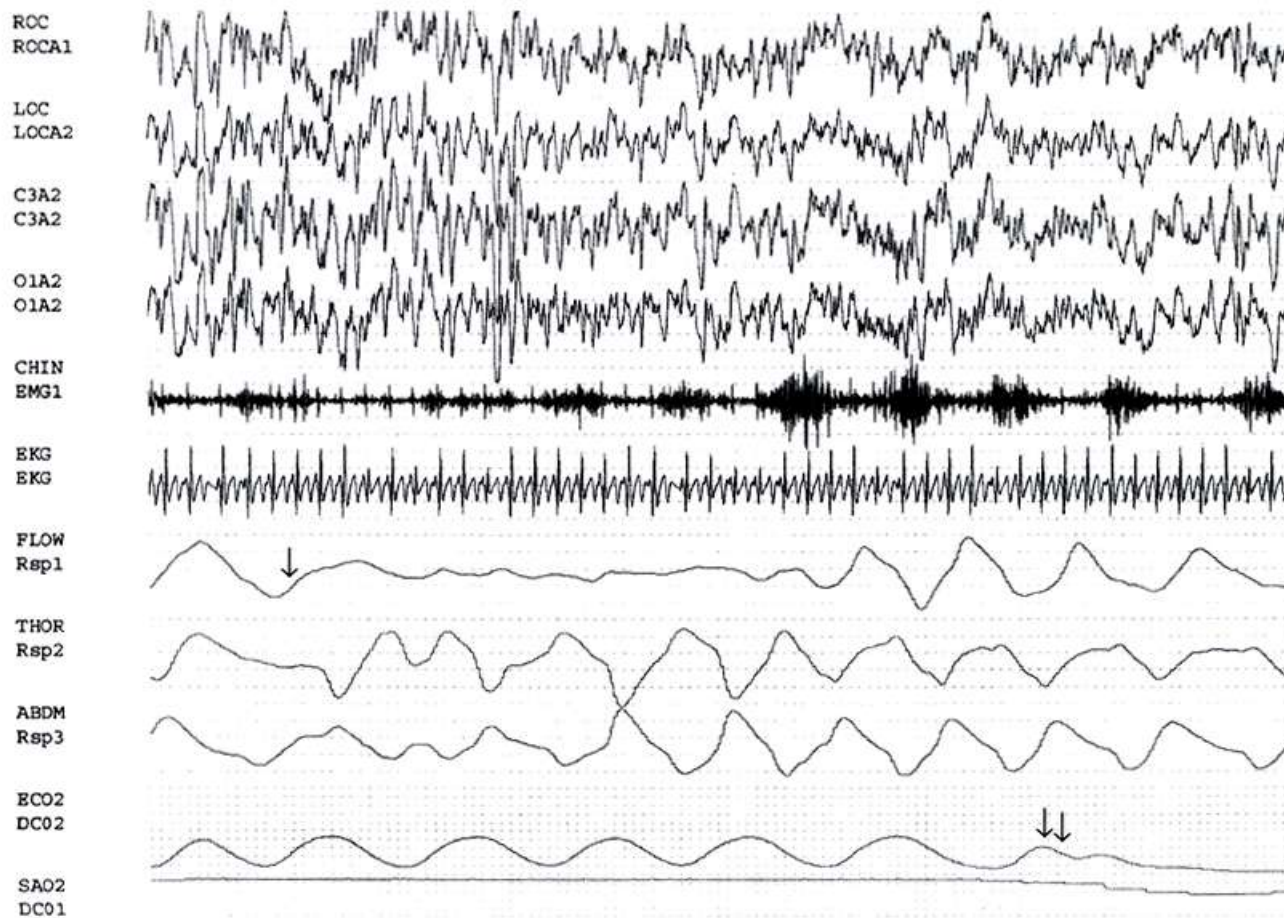


FIG. 14.2. Obstructive sleep apnea. ↓, initiation apneic event; ↓↓, oxygen resaturation.

Laboratory Evaluation

- Imaging of the Airway
 - Cine MRI
 - 3-D CT of the airway
 - Dental/oro-maxillofacial films
- Sleep video-endoscopy: under a general anesthesia that mimics sleep stage, directly visualize the airway in a dynamic state
- Microlaryngoscopy (static images)

Treatment Options I

- Treat Underlying medical conditions—
thyroid, reflux
- Weight loss
- Exercise
- Practice good sleep hygiene—no eating
before bed
- CPAP—Continuous Positive Airway Pressure

CASE STUDY--RJ

- 33 year old woman w/DS
- Essentially non-verbal, no significant signing, flat affect, morbidly obese, severe MR
- Treated for: Hypothyroidism, Hearing Loss Depression
- Moved to a group home—snoring noted
- PSG—severe OSA
- Started CPAP w/excellent compliance
- Interacting, talking, social

Treatment Options II

Relieve the Obstruction

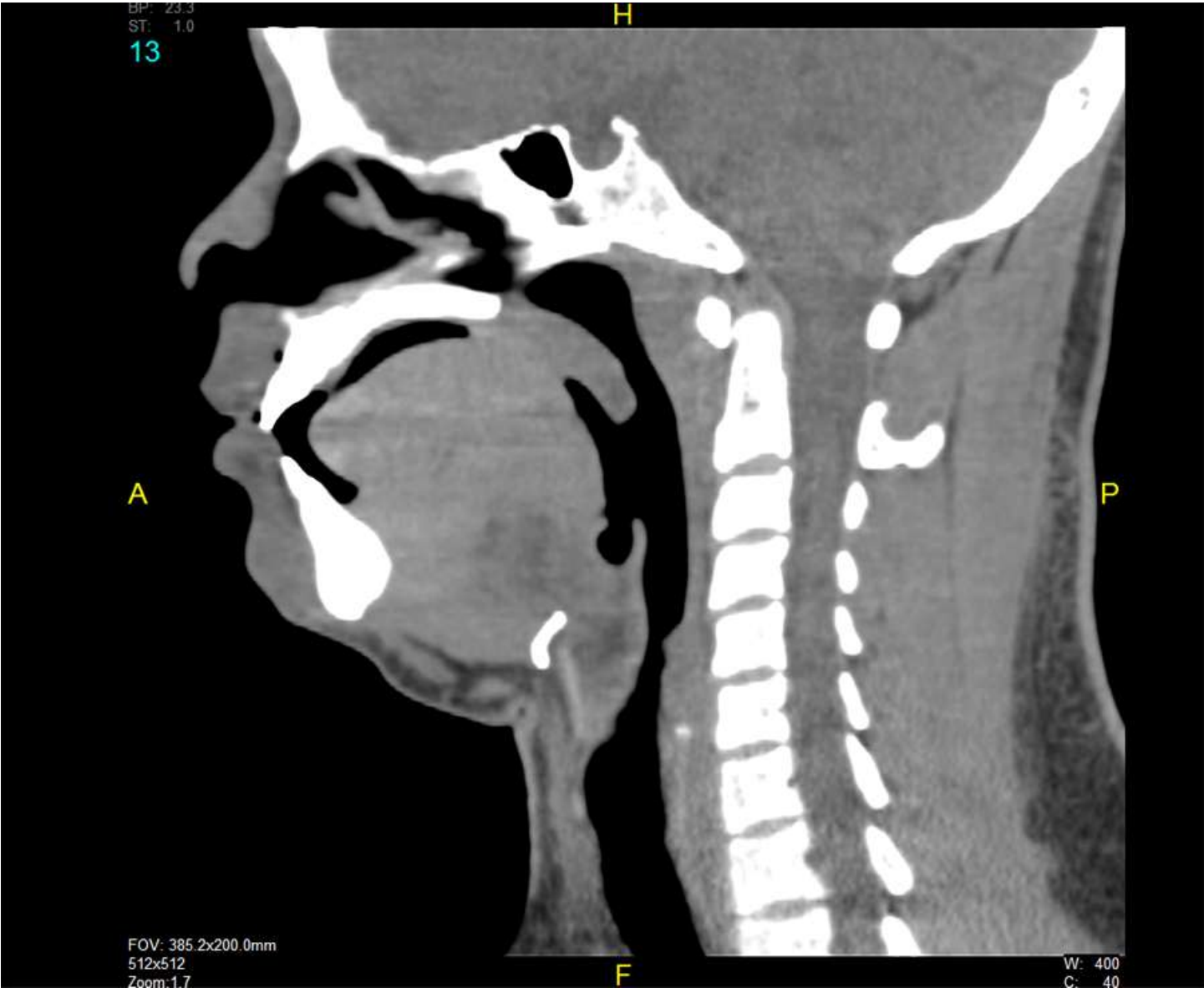
1. Tonsillectomy and Adenoidectomy
2. Orthodontic Appliances/Manipulation
3. Tongue retractors
4. Other surgeries:
 1. Tongue reduction
 2. Palatopharyngoplasty
 3. Hyoid Suspension/tongue mobilization
 4. Tracheotomy

CASE STUDY—GE

- 18 yo male w/DS
- Cc: Lack of energy, regression in interactions
- Hypothyroid, Asthma, EERD, Obesity, Diabetes, Moderate-severe MR
- Surgery: T&A age 6
- PSG: Severe OSA RDI=32.9, O2 desat.
- Endoscopy
- 3D Airway CT

Lingual Tonsil Hypertrophy





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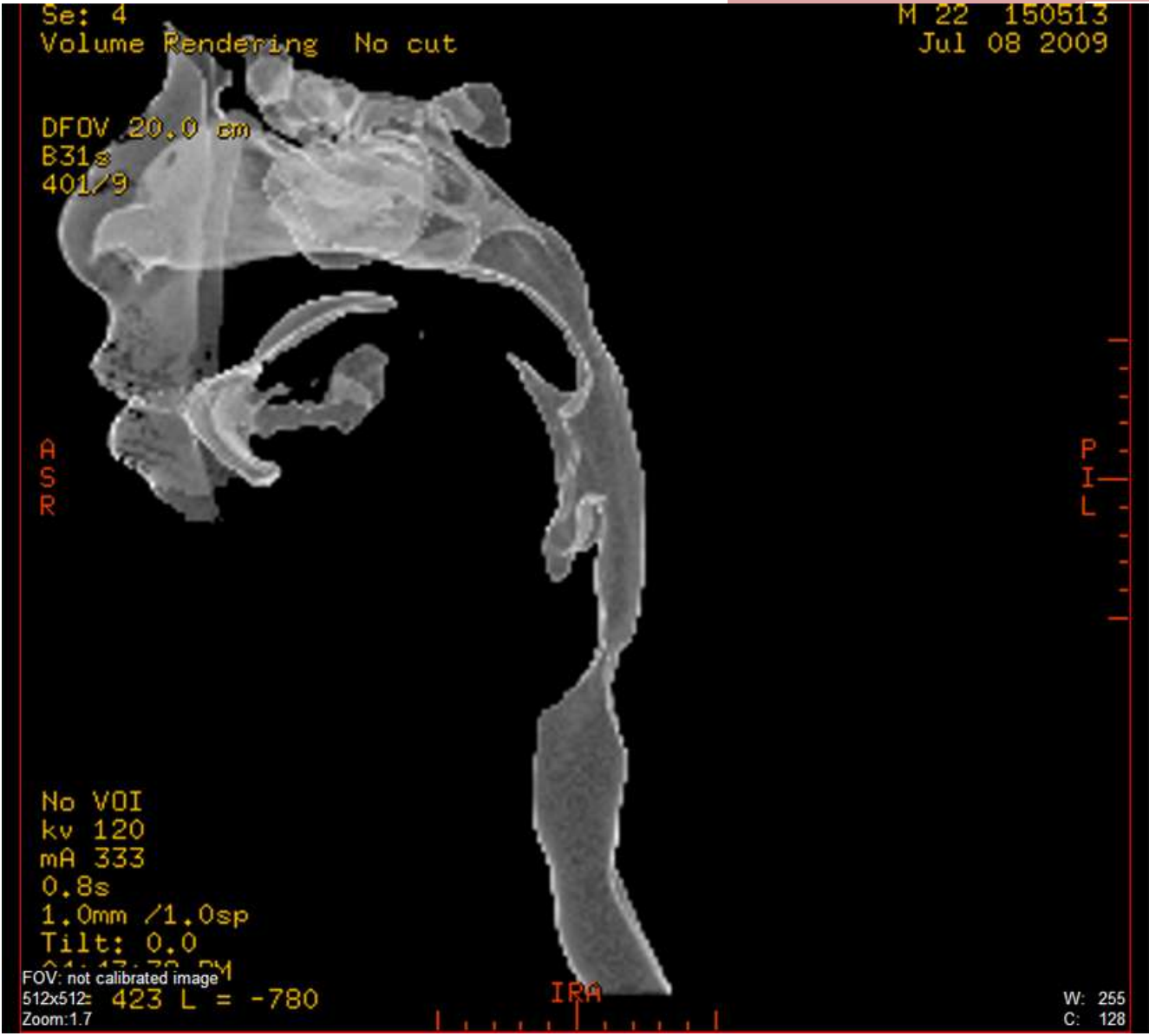
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CASE STUDY GE (cont.)

- 2008 Lingual tonsillectomy/Revision adenoidectomy/Tongue Base reduction
- Weight loss 30 pounds/exercise program
- Aggressive reflux treatment
- Continued sleep apnea but much better
- Not tolerate CPAP
- 12/2009 Supraglottoplasty, U3P
- Excellent clinical result but PSG still abnormal
- Able to use CPAP

Treatment Options II

Relieve the Obstruction

1. Tonsillectomy and Adenoidectomy
2. Orthodontic Appliances
3. Tongue retractors
4. Other surgeries:
 1. Tongue reduction
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Fig. 38.9. Adequate suspension creates a palpable dimple in the base of the tongue and just lifts the base of the tongue off the posterior hypopharyngeal wall. (Used with the express written permission of Influent Medical)

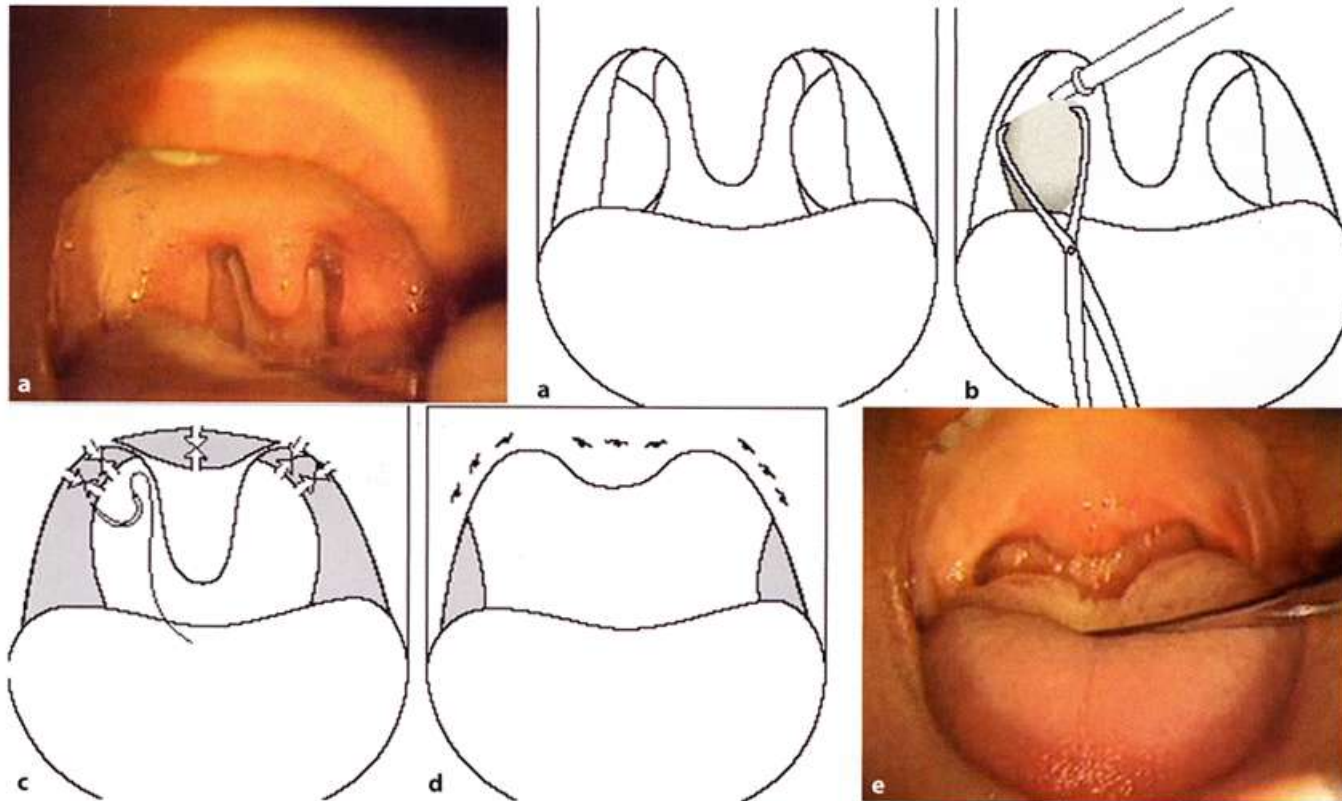


Fig. 43.5. Conservative UPPP. **a** Preoperative status in a 12-year-old boy with severe obstructive sleep apnea due to muscular hypotonia during sleep. Hypersomnia, hyperactive, loss of concentration, poor school performance. **b** Full tonsillectomy. **c** Removal of mucosa in fossa supratonsillar and an

oval incision of mucosa over the uvula. **d** Deep sutures in fossa supratonsillar as well as over the uvula. **e** *End result:* Note the fibrosis where the sutures were placed – normal sleep and alertness, improved grades in school

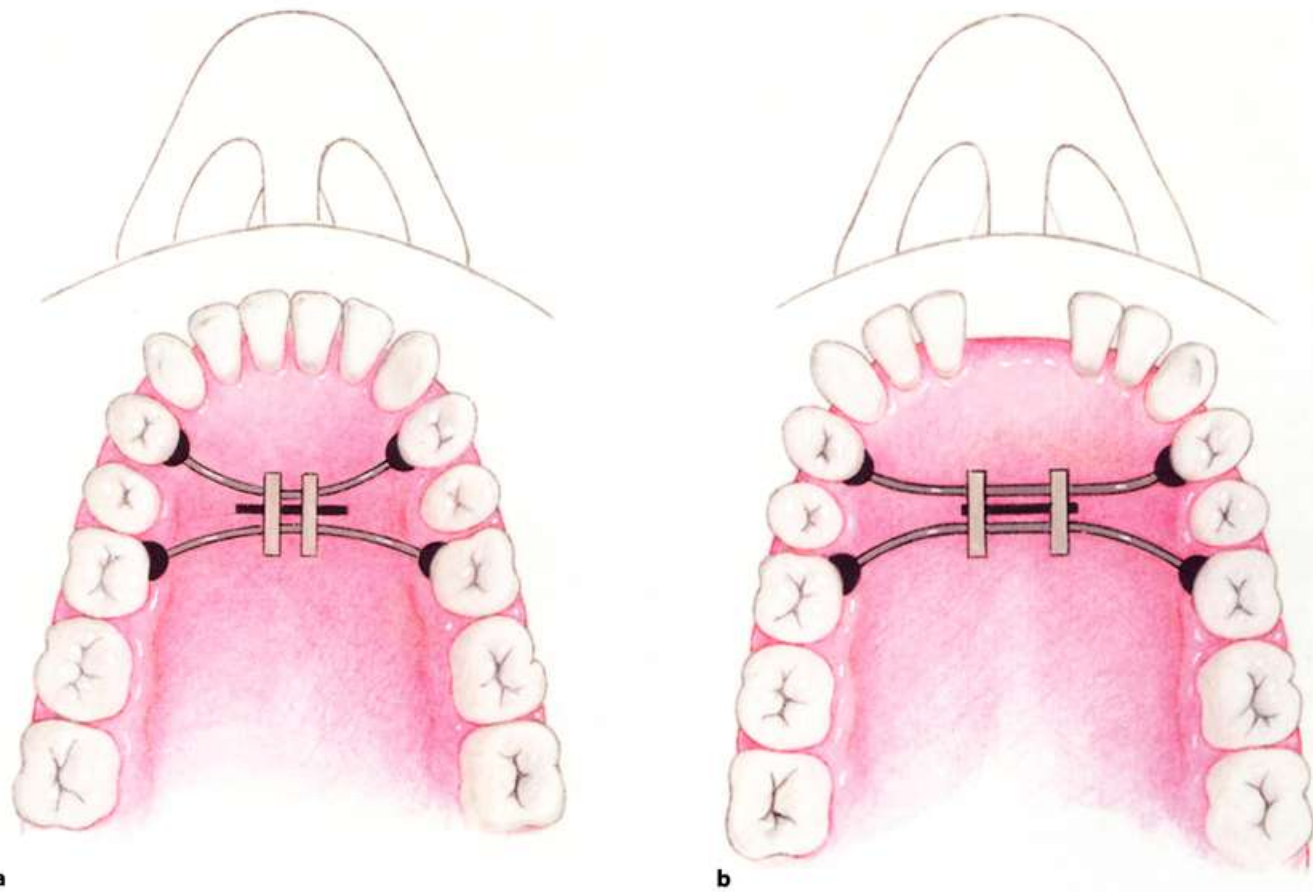


Fig. 42.3. Before (a) and after (b) maxillary widening

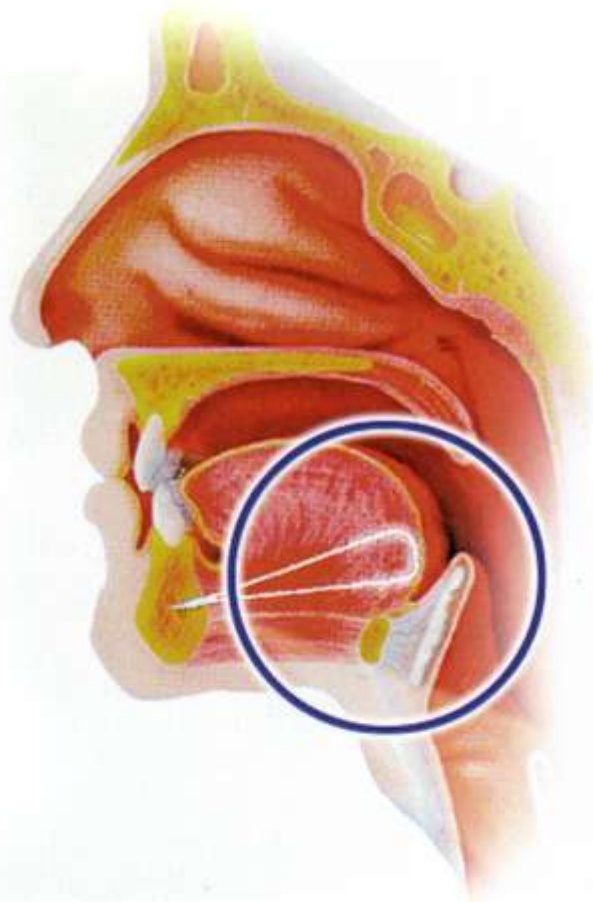


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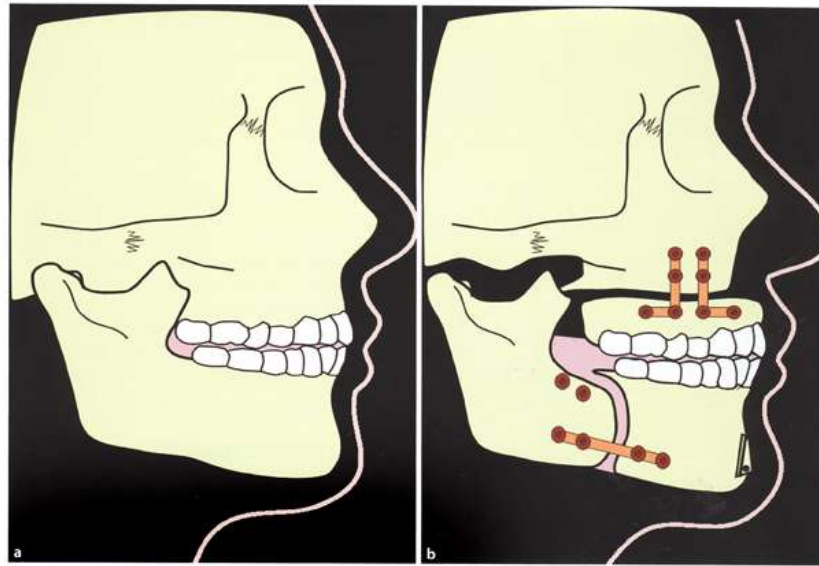


Fig. 41.3. Maxillomandibular advancement. a Before surgery, b After surgery



Fig. 41.4. Maxillomandibular advancement. a Preoperative cephalometric radiograph, b Postoperative cephalometric radiograph

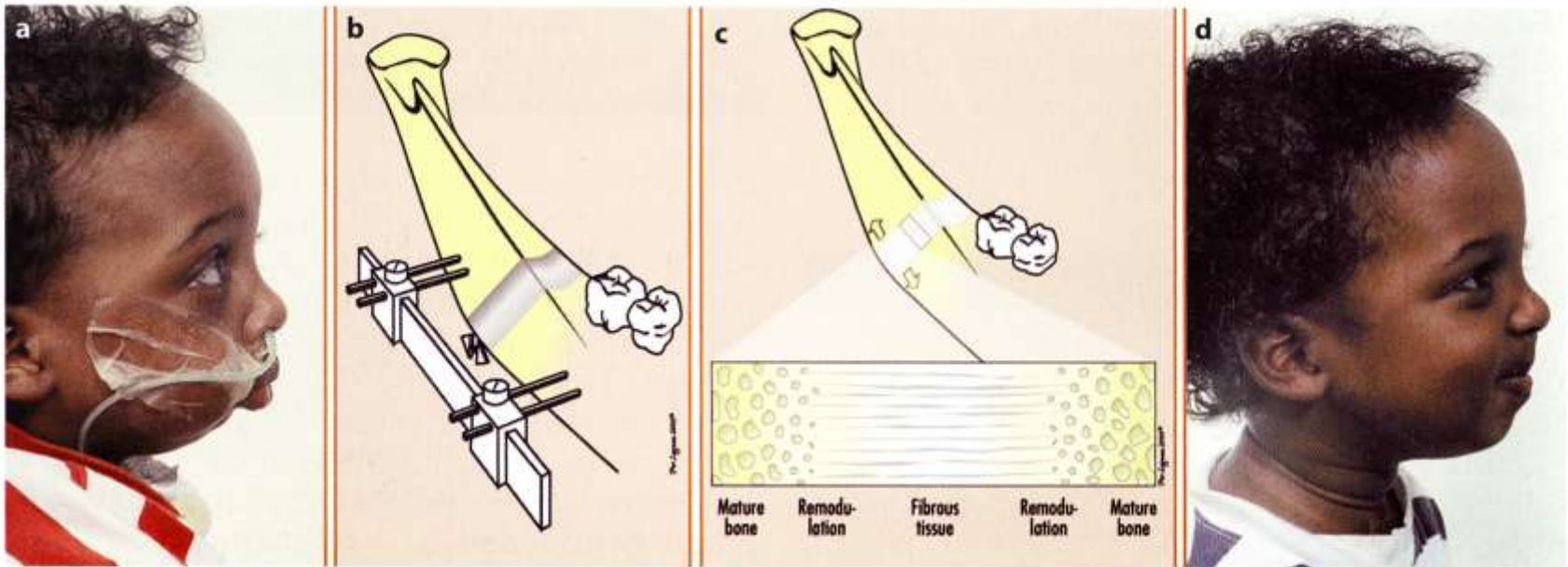


Fig. 43.2. A 2-year-old boy with severe sleep apnea **a** Mandibular hypoplasia, due to purulent infection in mandibular joints. **b** Distraction treatment with 1 mm widening each day.

c New bone is formed in the “artificial” fracture. **d** The boy 4 months later – decannulated and symptom-free



Thank you for your attention!

Any Questions?

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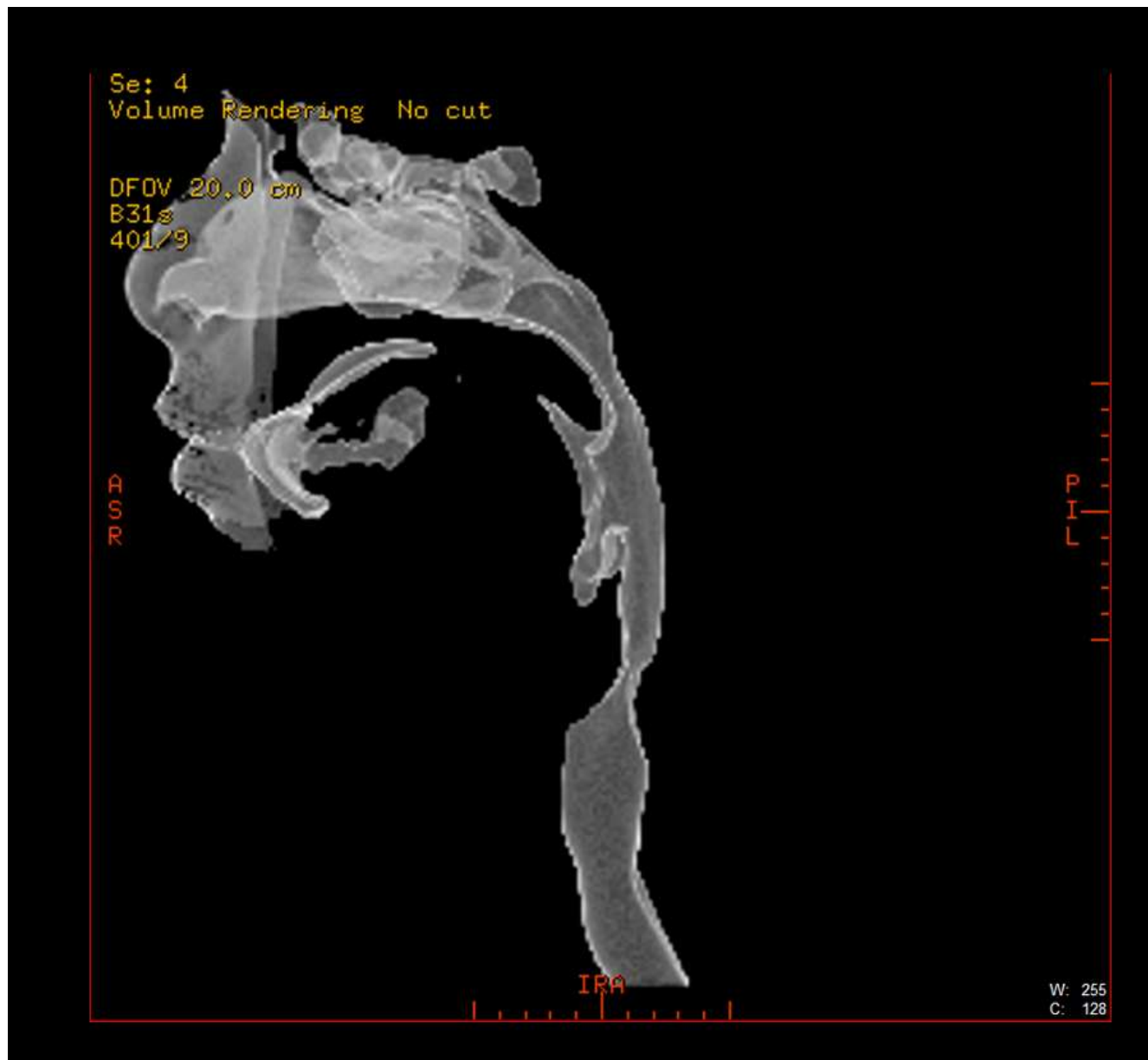
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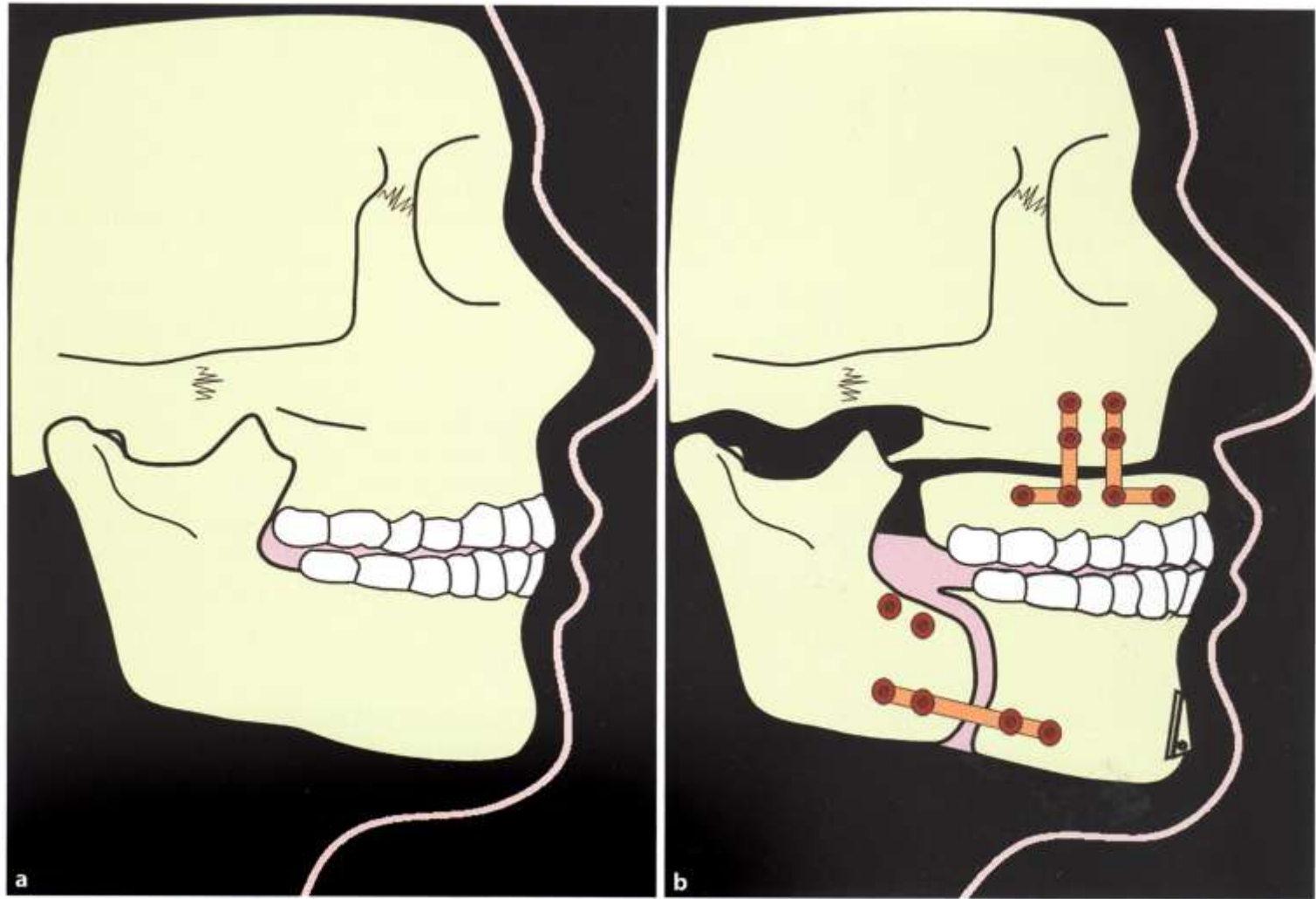


Fig. 41.3. Maxillomandibular advancement. **a** Before surgery. **b** After surgery

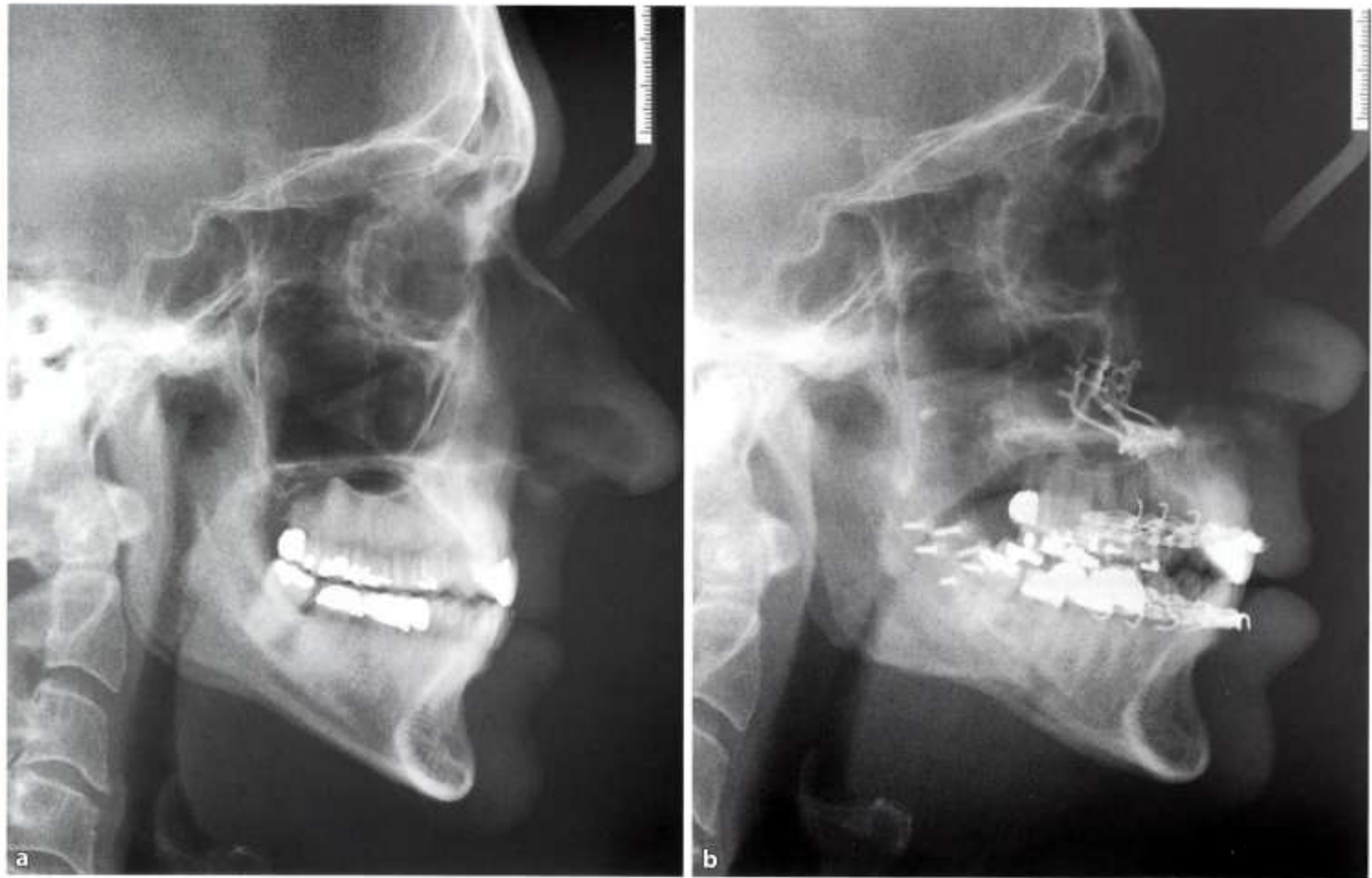


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