

大疱性表皮松解症的口腔表现和处理

原文地址：

http://www.debra.org/2006_PCC/Documents/twright-oralmanif.pdf

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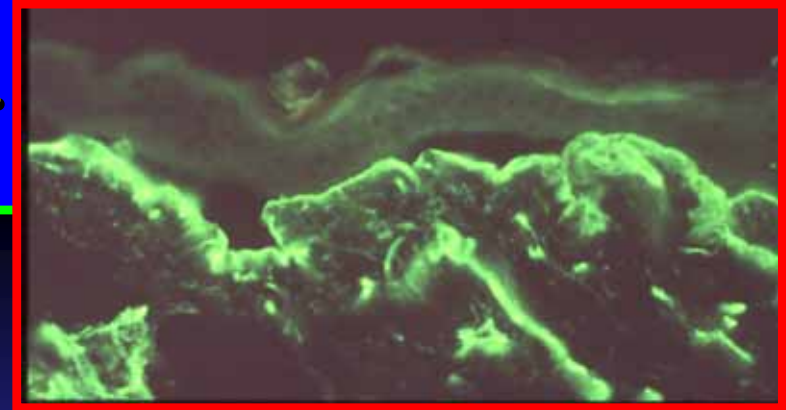
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因EB类型，分子缺陷，组织分离部位和其它因素的不同，口腔的表现可以有很大差异

Oral manifestations vary markedly depending on the specific EB type, the molecular defect, site of tissue separation and other factors.



- Simplex
 - Keratin genes – KRT5, KRT14
- Junctional
 - Laminin genes – LAMA3, LAMB3, LAMC2
- Dystrophic
 - Type VII Collagen – COL7A1 (over 600 mutations)

单纯型 -- 角蛋白基因 - KRT5 , KRT14

交界型 -- 层粘连蛋白基因 - LAMA3 , LAMB3 , LAMC2

营养不良型 -- 7型胶原 - COL7A1 (超过600种突变)

Oral Soft Tissue Manifestations of

EB EB的口腔软组织表现

- Highly variable depending on EB type
 - Increased fragility
 - Perioral lesions
 - Microstomia
 - Ulceration of mucosa, lips, cheeks, tongue, palate
 - Ankyloglossia
 - Obliteration of oral vestibule



与EB亚型高度相关

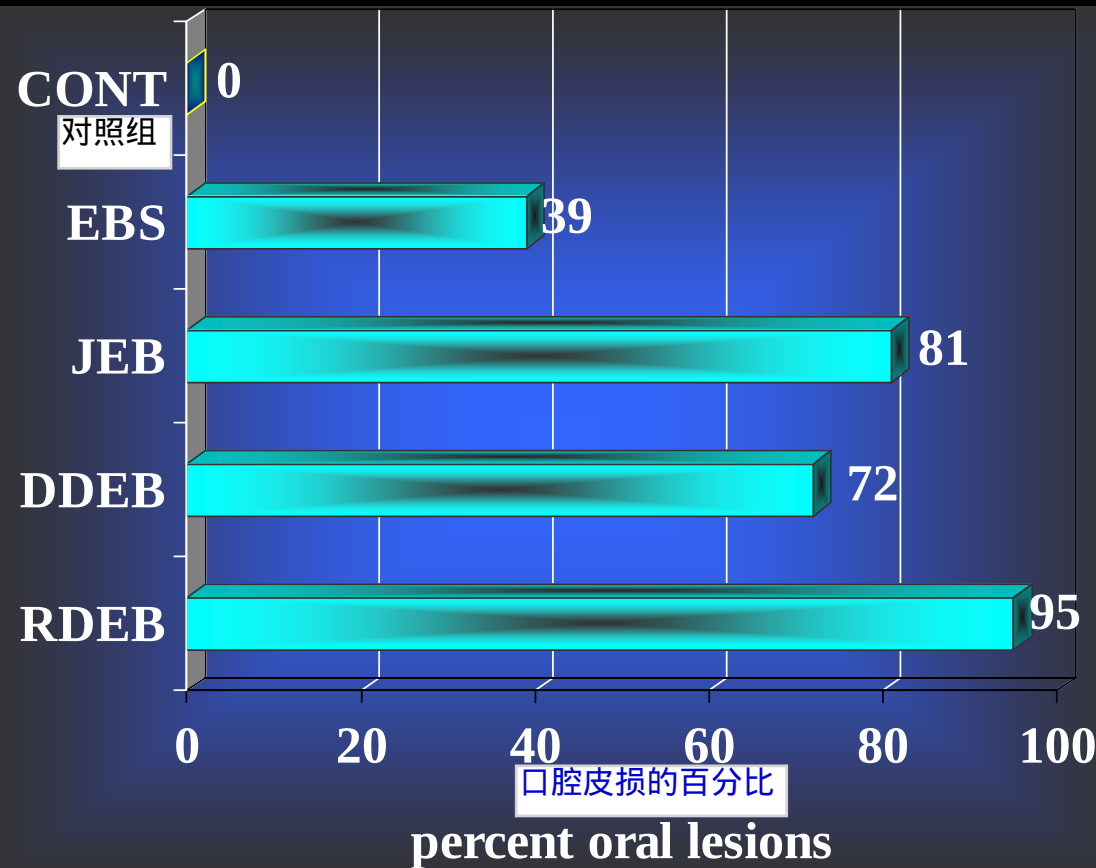
- 组织脆弱
- 口周皮损
- 小口
- 粘膜，嘴唇，面颊，舌头，上腭溃疡
- 舌粘连
- 无口腔前庭

EB Associated Soft Tissue Manifestations

- Can markedly affect ability to care for teeth.
- Can make providing oral health care extremely challenging.

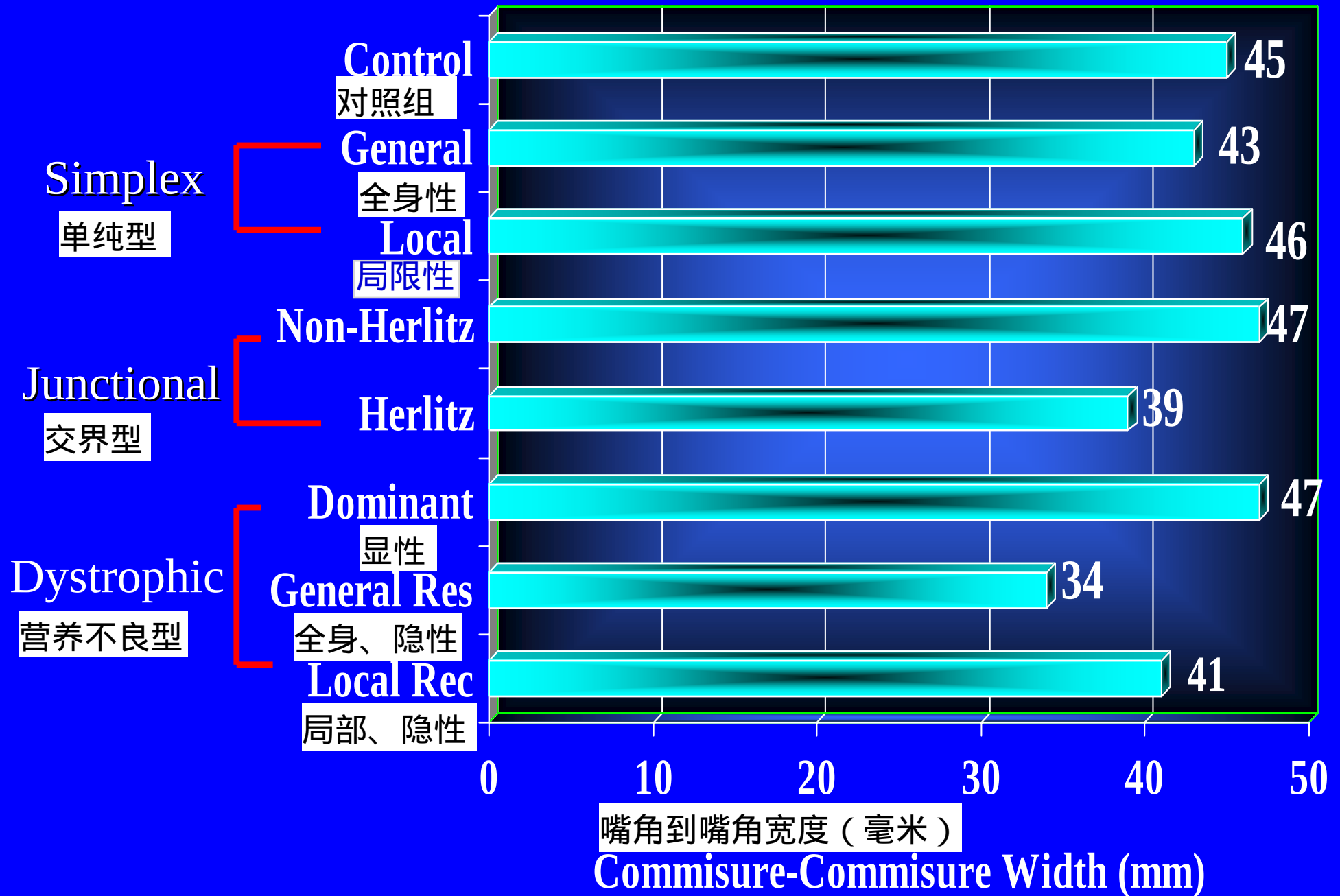
-- 显著影响牙齿护理的能力

-- 使口腔护理特别困难



不同EB类型的口腔开口宽度

Oral Cavity Width in Different EB Types



Oral Soft Tissue Screening

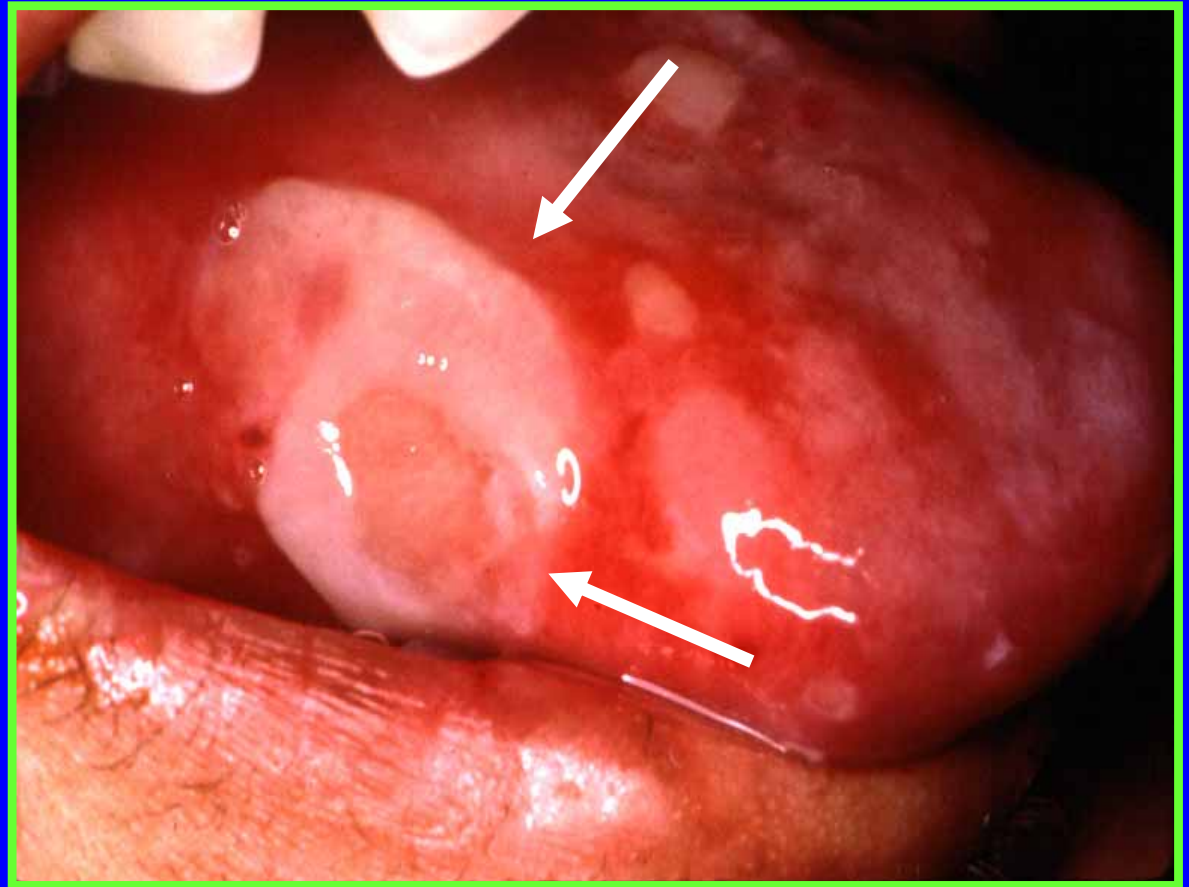
- Individuals with Recessive Dystrophic EB are at increased risk for oral carcinoma.
- Self examination – monitor oral lesions for marked changes, especially lesions developing hard or rolled borders and increasing in size.
- Professional oral examination every 6 months

RDEB患者得口腔癌症的风险比普通人高。

自我检查 -- 观察口腔皮损有无明显改变，特别是伤口外缘变硬或隆起，以及范围变大。
每6个月做正规的口腔检查

口腔软组织检查

Oral Soft Tissue Screening



处理不连续的软组织损伤

Managing Discrete Soft Tissue Lesions



Cyanoacrylate based topical covering that provides relief from sensitive oral ulcers.

基于氰基丙烯酸胶的外用覆盖物，可以缓解口腔溃疡的疼痛

适用于口腔炎症的漱口水

Mouthwash for Stomatitis



- Treat stomatitis palliatively in the absence of infection
- Magic Mouthwash
 - Maalox (100ml)
 - Viscous Xylocaine (25-50ml)
 - Benadryl (25-50ml)

没有感染的时候温和的处理口腔炎
魔力漱口水

- 氢氧化铝、氢氧化镁混合物 (100毫升)
- 苦息乐卡因液 (25 ~ 50毫升)
- 苯那君 (25 ~ 50毫升)

口腔念珠菌病（鹅口疮）

Oral Candidiasis

- Individuals with chronic oral ulcerations (e.g. RDEB, Dowling Mera Simplex) are at increased risk for oral candida infections (thrush).
- Typically treated with oral nystatin or ketoconazol.
- Treatment with chlorhexidine rinse can reduce candida overgrowth.

-- 慢性口腔溃疡患者（RDEB，EBS-DM）患口腔念珠菌病（鹅口疮）的可能性高。
-- 通常用制霉菌素或酮康唑治疗
-- 用洗必太漱口水可以抑制真菌生长

Dental Problems and EB

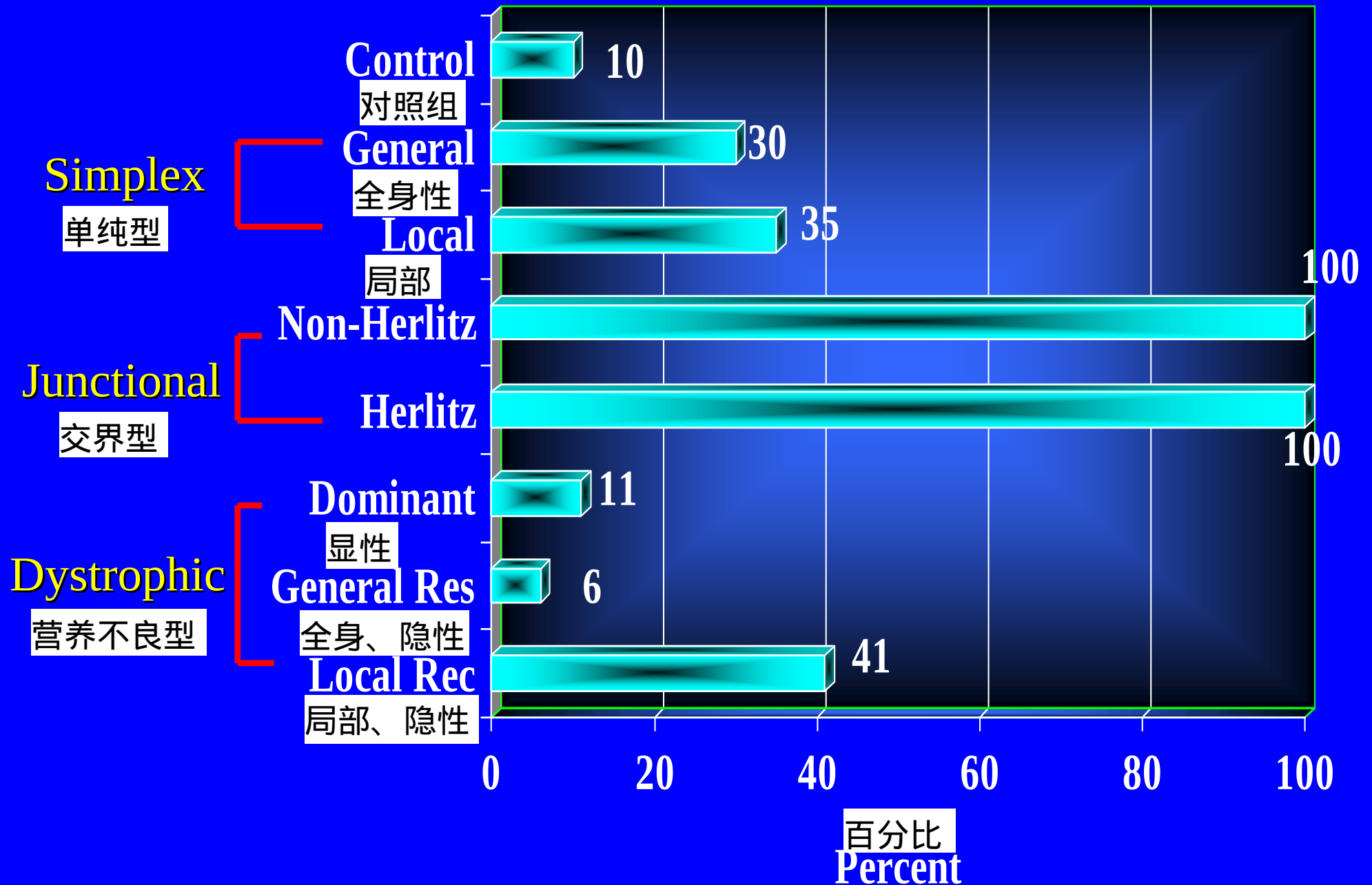


- Junctional EB and occasionally other EB types will have severe enamel hypoplasia.
- Individuals with junctional and recessive dystrophic EB are at increased risk for dental caries.
- Individuals with recessive dystrophic EB frequently have severe dental crowding and a deep bite.

-- 交界型EB，和少数其它EB类型的人会发生严重的牙釉质发育不全
-- RDEB和交界型EB患者患龋齿的风险较高
-- RDEB患者经常有严重的牙列拥挤和深覆牙合

不同EB类型中的牙釉质发育不全

Enamel Hypoplasia in Different EB Types



交界型大疱性表皮松解症

Junctional Epidermolysis Bullosa

层粘连蛋白缺陷影响皮肤和牙齿形成

Laminin defect affecting skin and tooth formation.



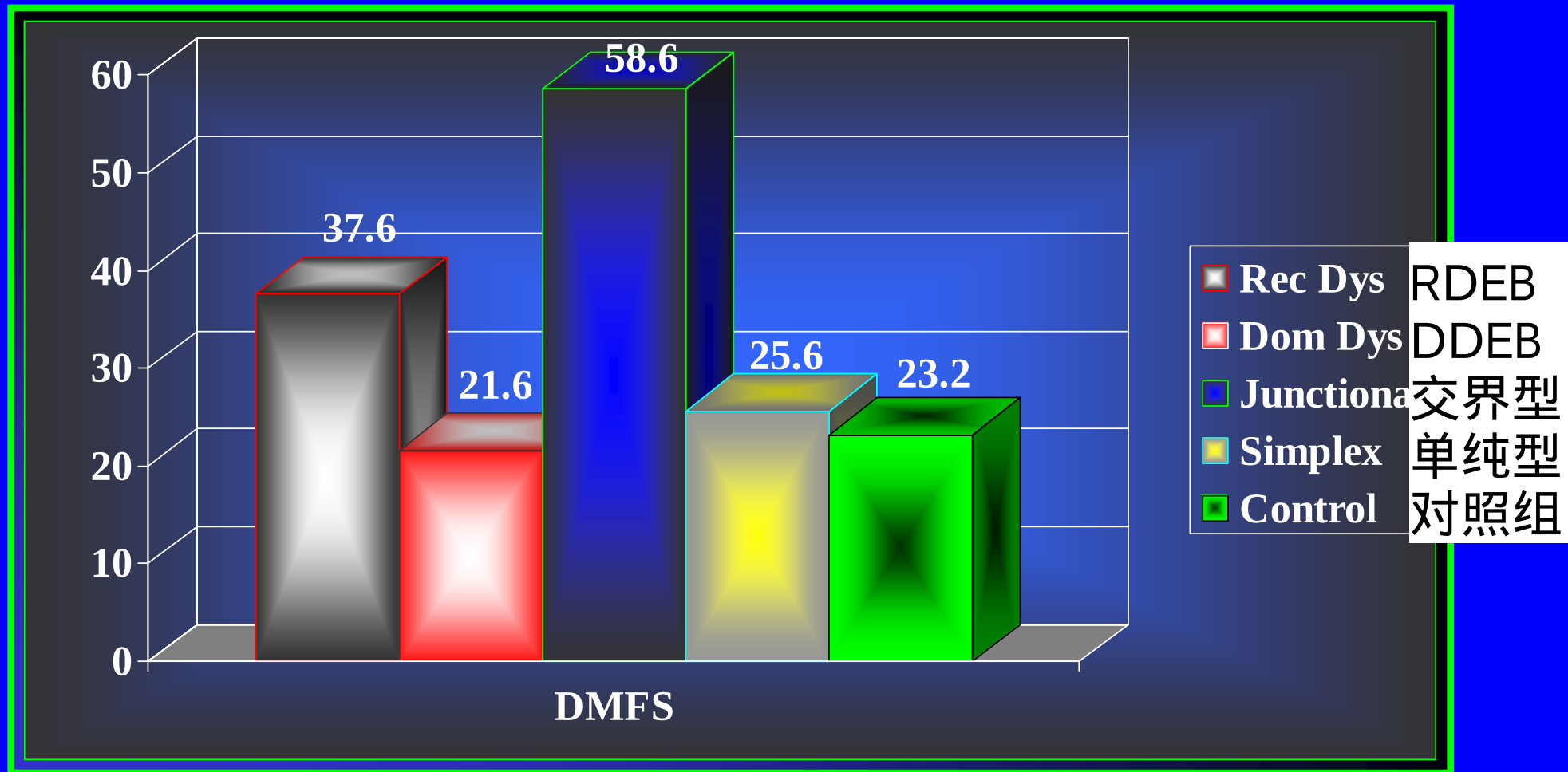
Factors Contributing to Increased Dental Caries Risk in EB

- Enamel hypoplasia
- Consumption of high calorie (carbohydrate) diet
- Slow eaters and increased feeding frequency
- Reduced oral clearance of food due to soft tissue strictures
- Inability to effectively brush teeth
- Difficulty receiving routine preventive dental care.

- 牙釉质发育不全
- 高热量饮食（碳水化合物）
- 吃饭慢，饮食频率高
- 由软组织损伤引起的口腔护理减少
- 不能有效的刷牙
- 不能接受有规律的牙科预防治疗

Prevalence of Dental Caries in EB Population

EB患者群中的牙齿护理普及率



decayed,missing or filled surfaces 龋失补牙

Caries Prevention

- Diet Control
 - Minimize frequency of exposure to refined carbohydrates
- Mechanical plaque/substrate control
 - Brushing, flossing, rinsing
- Chemotherapeutic control
 - Fluoride, chlorhexidine
- Protective covering of teeth
 - Sealants, crowns

控制饮食

-- 减少牙齿与精制碳水化合物接触频率

机械方法控制牙菌斑

-- 刷牙，牙线，漱口

化学方法控制

-- 氟化物，洗必太

预防性补牙

- 封牙，假齿冠



牙刷选择

Toothbrush Selection

嘴张不开、口腔粘膜或牙龈非常脆弱的人

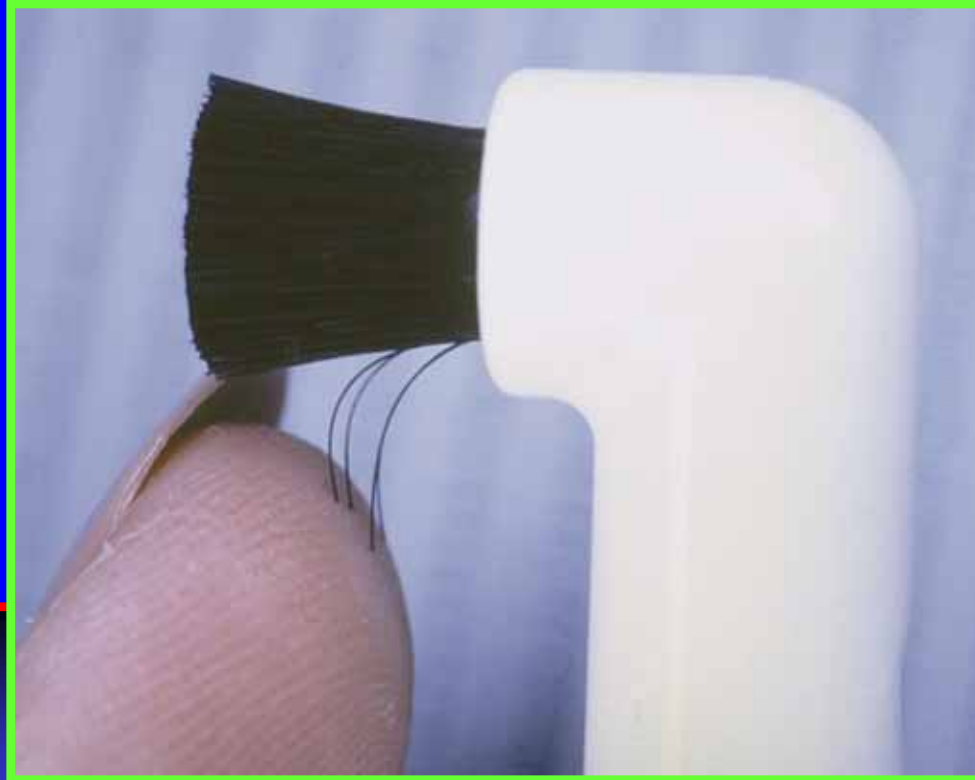
- 选择头小的牙刷
- 毛要软
- 用热水泡一下牙刷，使毛更软
- 考虑使用小头、软毛的电动牙刷



- Individuals with microstomia or severe fragility of the oral mucosa and gingiva.
 - Select small headed brush
 - Select soft bristles
 - Run brush under hot water to further soften bristles
 - Consider mechanical brush with small head and soft bristles.

电动牙刷

Mechanical Toothbrush



RotoDent
Pro-Dentec
PO Box 3889
Batesville, AR 72503
800-228-5595



Chemotherapeutic Caries Control

- Fluoride therapy
 - Systemic – H₂O, Supplements
 - Topical - Toothpaste, Mouthwash
 - Professional Application – Gel, Varnish
- Antimicrobial Agents
 - Chlorhexidine

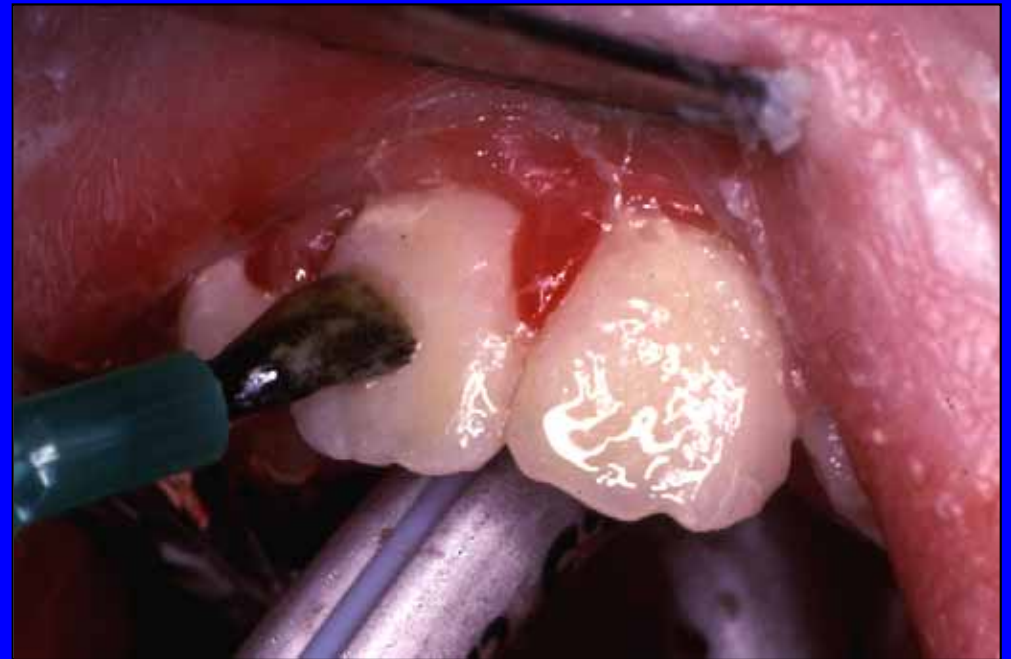


氟化物疗法

- 全身性 - 水中添加
 - 外用 - 含氟牙膏、漱口水
 - -- 专业护理 - 凝胶，氟化物涂膜
- 抗菌剂
- 洗必太

氟化物涂膜

Fluoride Varnish Placement

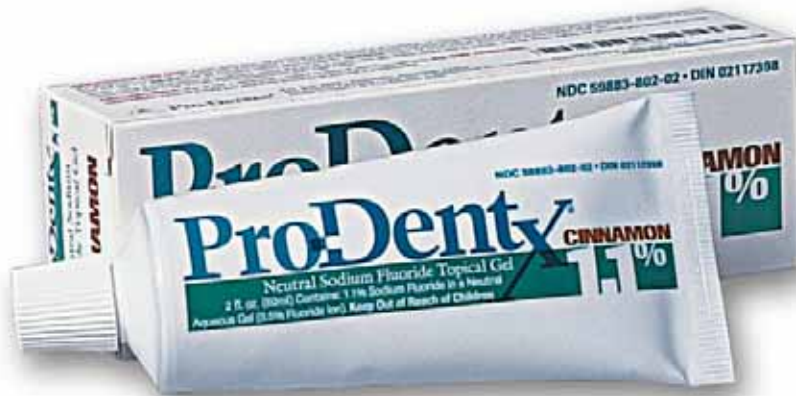


高氟处方牙膏

Fluoride Concentration of Rx Toothpastes

- Control RX, Prevident (NaF) = 0.5% F 5000ppm
- 5X stronger than regular toothpaste
- Rx only

Control Rx (商标) , Prevident (商品名) , 含0.5%氟化钠
含氟比普通牙膏多5倍以上
处方药



ControlRx™ - 5000 ppm F, 1.1% NaF Dentifrice

ControlRx 5000 ppm prescription fluoride therapy can be used in place of regular toothpaste for daily cleaning. Plus, new ControlRx is the only neutral sodium fluoride dentifrice that contains Microdent®, a patented formula for controlling plaque. Microdent reduces the Surface Free Energy of tooth surfaces[®], making it more difficult for plaque to adhere.



- ▲ Caries Reduction
- ▲ Recommended for Relief from Sensitivity
- ▲ Protection from Root Caries
- ▲ Remineralization
- ▲ Refreshing Vanilla Mint Flavor

洗必太漱口水

Chlorhexidine Rinse

- Effective antimicrobial rinse
- Reduces candidiasis
- Reduces dental caries

有效的抗菌漱口水
杀灭念珠菌
减少龋齿



EB患者的牙齿修补

Dental Restorations for EB Patients



- If extensive soft tissue and dental caries problems then - Use techniques and materials that cover the teeth.

- Stainless steel crowns
- Resin bonded stainless steel crowns
- Porcelain fused to metal crowns
- Ceramic or new tooth colored crown materials

如果软组织和龋齿问题严重，应用修补材料和技术

- 不锈钢牙冠
- 树脂粘固不锈钢冠
- 金属烤瓷冠
- 全瓷牙或其它接近牙齿本色的材料

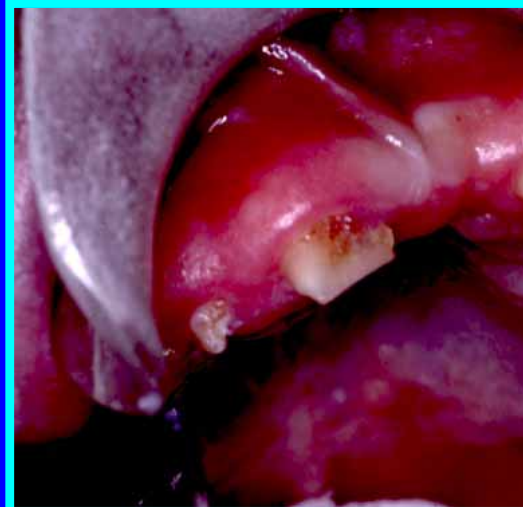
交界型EB患者的乳牙和早期恒牙牙冠

Crowns for Primary and Early Permanent Teeth – Junctional EB



青少年恒牙齿系完全复原 --交界型EB

Complete Restoration Young Permanent Dentition – Junctional EB



全烤瓷牙/金属烤瓷牙 -- 交界型EB

Full Mouth Porcelain/Metal Crowns Junctional EB



EB患者种牙

Dental Implants in EB



Implants in patient
with RDEB- Inversa

RDEB-Inversa患者种牙

种牙时要考虑的因素

Dental Implant Considerations

- Oral opening access
- Bone available to receive implant
- Soft tissue health
- Ability to maintain implants
 - Oral hygiene

嘴开口大小
牙床是否还在，能否接受种植
软组织是否健康
种植的牙能否成活
-- 口腔卫生



RDEB患者种牙

Dental Implants in RDEB Patient



Dental Malocclusion is Prevalent in RDEB Due to Soft Tissue Constriction and Crowding

牙齿咬合不正在RDEB患者中很常见，原因是软组织收缩和牙齿拥挤

- Can allow better alignment of incisors by selectively extracting primary canines and permanent first premolars.

选择性拔掉乳牙犬齿和恒牙第一白齿可以使门牙排列更整齐



牙齿护理

Providing Dental Treatment



- In mildly affected individuals dental treatment can normally be provided with minor modifications to approach.
- Severely affected individuals will often require treatment with the aid of general anesthesia.

正常的程序稍微修改一下就可以给轻微的患者做牙齿护理
严重的患者在护理牙齿前需要麻醉

Indications for General Anesthesia to Provide Treatment in EB Patients

- Extensive soft tissue fragility
 - Oral blistering and scarring
 - Limited oral access due to microstomia
- Rampant dental caries
 - Extensive restorative or surgical Tx required
- Invasive procedures
 - Implant placement
 - Soft tissue surgery



大面积软组织破损
-- 口腔水疱和疤痕
-- 由小口造成的不易深入口腔
剧烈的牙齿护理
-- 大面积的修复或手术
侵入性的手术
-- 植牙
-- 软组织手术

EB和全身麻醉

General Anesthesia and EB

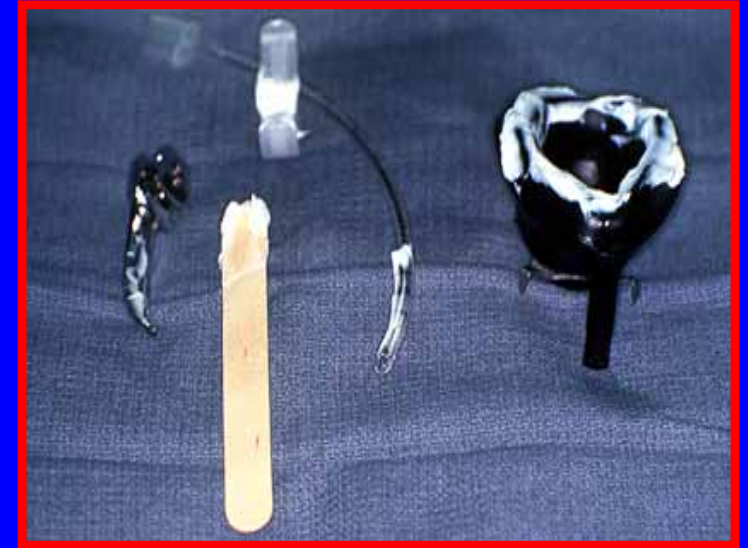


- Allows comprehensive treatment of dental caries.
- Following special protocol greatly reduces soft tissue damage from procedure.
- Safety record based on literature of hundreds of cases using endotracheal intubation is excellent.

可以全面的处理龋齿
按照特殊的流程，大大减少软组织伤害
根据已有的数百例气管插管报道，安全性良好

EB和全身麻醉

General Anesthesia and EB



- Use endotracheal intubation
- Use atraumatic techniques
- Lubricate instruments and tissues to prevent shear forces
- No tape or adhesives

气管内插管
使用减少损伤的技术
润滑器械和组织，防止剪切力
不用胶带

Treatment and Vital Sign Monitoring Without Adhesives

- Pulse oximetry monitor
 - Block adhesive on probe
 - Clip probe
- EKG leads
 - Use non-adhesive leads
 - Cut adhesive off leads
 - Use patient's weight to make contact



脉搏和血氧监控

-- 遮住探针上的粘胶

-- 用夹子固定探针

心电图的引脚

-- 使用没有粘性的引脚

-- 把粘性引脚切掉

-- 靠患者体重保持接触

Dental Management of EB

- Early intervention and dental consult
- Regular evaluation of soft tissues for premalignant alterations
- Aggressive prevention approaches
- Regular recall appointments
- Restorative therapy provided before advanced destruction of the teeth



尽早开始护理和治疗

周期性检查软组织，在恶化前矫正
主动预防

周期性到医院检查

在牙齿严重损伤之前进行补救

现代的预防和治疗技术已经可以使
口腔症状严重的EB患者保持外观良
好、功能正常的牙齿

esthetic and functional dentition:



