



European  
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# **Dental care pathways for patients with epidermolysis bullosa: consensus clinical position statement from the European Reference Network for Rare Skin Diseases**

Ge, Fr, It, Au, UK, The Neth

- Aim: To provide to guide for patients with each type of EB to seek advice at the different dental services.
- First draft: done
- Consensus: to be obtained
- Expression of interest > Cristina
- Send back: end of April 2023

## EB Simplex (keratin and KLHL24 mutations)

Oral ulcers in patients with localized and intermediate EBS due to keratin mutations, have been described in 7% to 35% of the patients.<sup>3,4</sup> In patients with intermediate EBS with cardiomyopathy due to mutations in KLHL24 the oral mucosa is mildly affected, with oral ulceration in 43% of the affected groups.<sup>5,6</sup> Severity tends to lessen with age.

### First referral

- General dentist according to national guidelines.

### Follow up:

- According to local (national) guidelines for the general population.

### Specialist:

- Oral Medicine Specialist or allied subject area/specialty: Once a year or as needed according to severity of the mucosal fragility.

## **Consensus agreement**

<b>Recommendation:</b>	<b>Do you agree?</b>	<b>What would you recommend in addition or instead?</b>
<u>First referral</u> General dentist according to national guidelines.	<b>Yes/No</b>	
<u>Follow up:</u> According to local (national) guidelines for the general population.	<b>Yes/No</b>	
<u>Specialist:</u> Oral Medicine Specialist or allied subject area/specialty: Once a year or as needed according to severity of the mucosal fragility.	<b>Yes/No</b>	

### First referral

- Paediatric dentist (if available, otherwise a general dentist) at the age of 3 to 6 months.

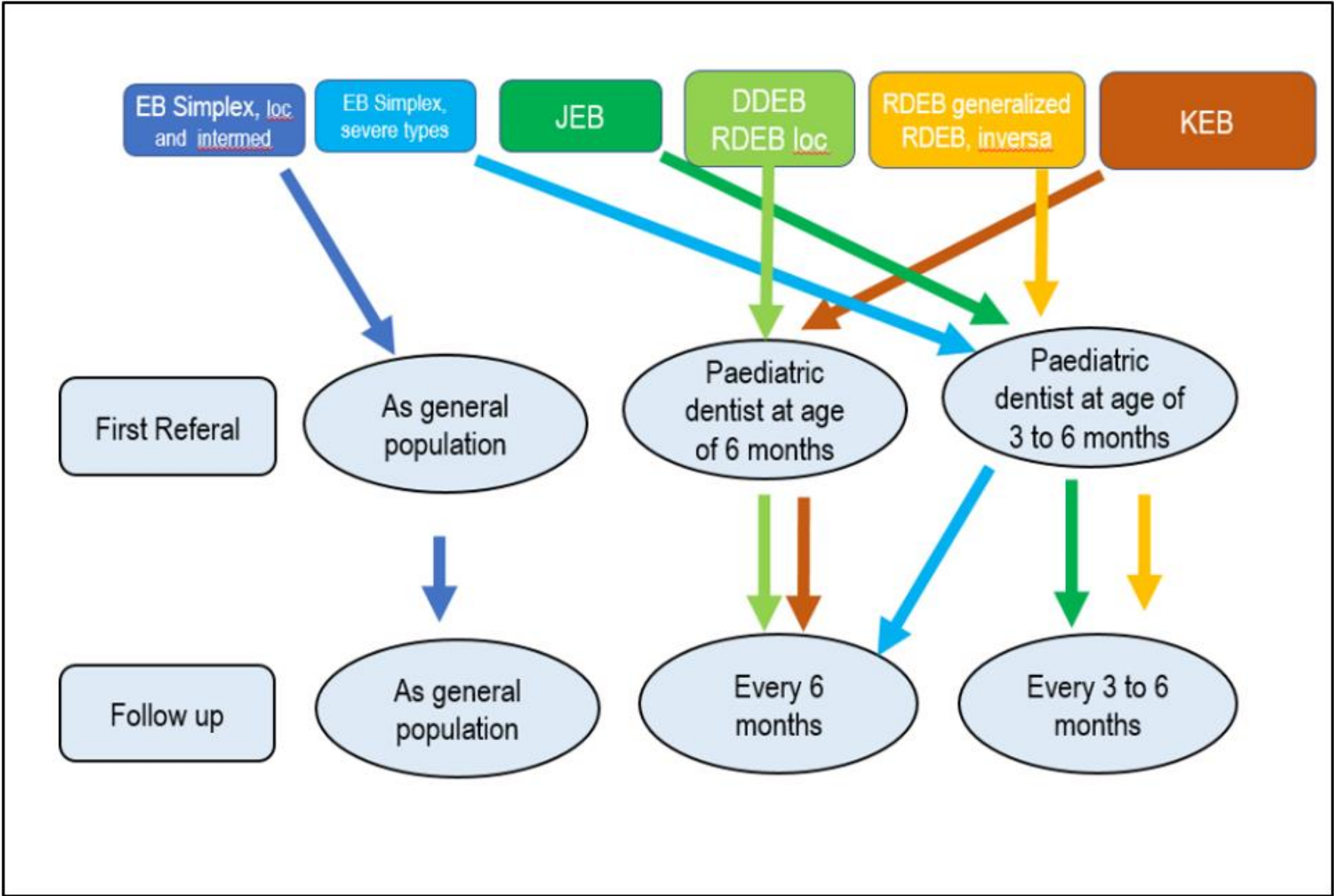
### Follow up:

- Every 3 to 6 months (frequency can vary according to specific needs)

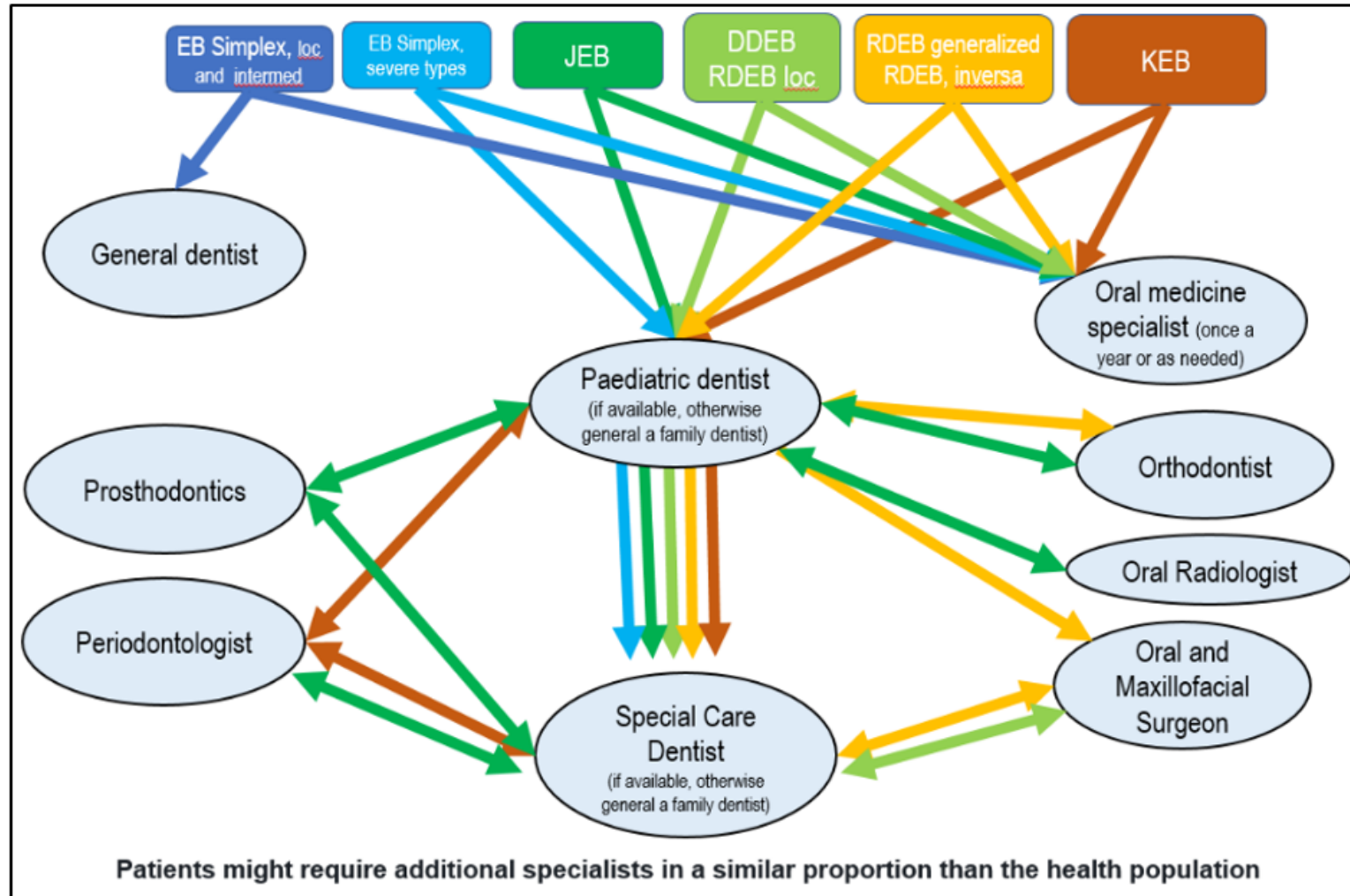
### Specialist:

- *Paediatric dentist* (if available, otherwise a general dentist) during temporary and mixed dentition, up to age of 12 to 18, depending on the country. Most patient will need extensive restorations or crowns. This can be provided either by a Paediatric dentist or a specialist in *Prosthodontics* or a general dentist if he/she is nearer to the patient's home.
  - *Regular Dental X rays* to check for crown resorption and tooth retention during the temporary and mixed dentition.
  - *Orthodontist* as part of the team during the maxillofacial growth and development.
  - *Specialist in Prosthodontics (or Oral Rehabilitation)* will assess the need for indirect restorations like veneers or crowns due to Amelogenesis Imperfecta. The age at which the specialist in Oral Rehabilitation provides the care will depend on local pathways. It needs to be well coordinated with the Paediatric Dentist and the Special Care Dentist.
  - *Special Care Dentist* (if available, otherwise a general dentist) takes over the overall care of the patient at the age of 12 to 18, depending on the country, and will continue the preventative strategies and leasing with other specialities as needed.
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- *Oral Medicine Specialist* or allied subject area/specialty: Once a year or as needed according to severity of the oral ulcers.

**Figure 1. Flow diagram: Dental care pathways for patients with EB: First referral and follow up.**



**Figure 2. Common referral Pathways in different types of EB.**



Note: If the different specialities are not available in the country, general dentist can also provide the treatment required.