


UNIVERSITÄT
KINDERSPITAL
ZÜRICH



Gastrostomy in EB



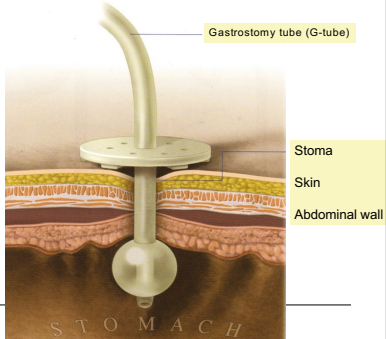
Agnes Schwiager-Briel
EB-KidZ
University Children's Hospital Zürich

Blau Spital der Elternvereinfachung

1

Gastrostomy

= permanent opening from the external surface of the abdominal wall into the stomach, usually for inserting a feeding tube.



Gastrostomy tube (G-tube)

Stoma
Skin
Abdominal wall

STOMACH

UNIVERSITÄT
KINDERSPITAL
ZÜRICH

2

Gastrostomy in EB-the main questions

WHO

WHEN

WHY

HOW

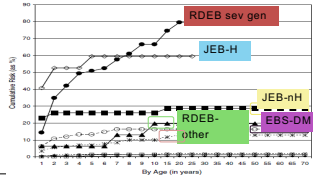


UNIVERSITÄT
KINDERSPITAL
ZÜRICH Schwiager-Briel, Gastrostomy in EB, EB-Clinet, Vienna 2024

3

WHO

Majority of EB patients with G-tube insertions have RDEB

Feinstein et al. JAMA 2019: 36.7 % of RDEB pts have a G-tube placement ; mean age 5.5 yrs (2-10.3 yrs)

Risk of growth retardation in EB (Fine 2008)

UNIVERSITÄT
KINDERSPITAL
ZÜRICH

4

WHO

Majority of EB patients with G-tube insertions have RDEB

Feinstein et al. JAMA 2019: 36.7 % of RDEB pts have a G-tube placement ; mean age 5.5 yrs (2-10.3 yrs)

Kleinman et al. 2016

- RDEB 75%
- JEB 15%
- EBS 10%

- G-tubes can be necessary in **any type of EB**
- decisions according to individual settings have to be made

Yang et al. Am J Clin Dermatol 2016: controversy of G-tubes in JEB severe gen is discussed



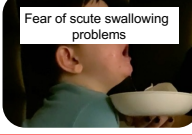
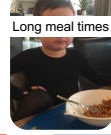
Marro et al. Orphanet J Rare Dis 2021: G-tubes in infants with EBS severe gen

- 9 infants, >25% skin involvement
- mucous membrane involvement and FTT

UNIVERSITÄT
KINDERSPITAL
ZÜRICH Schwiager-Briel, Gastrostomy in EB, EB-Clinet, Vienna 2024

5

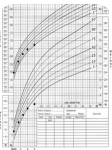


WHY









Fear of scute swallowing problems

Long meal times

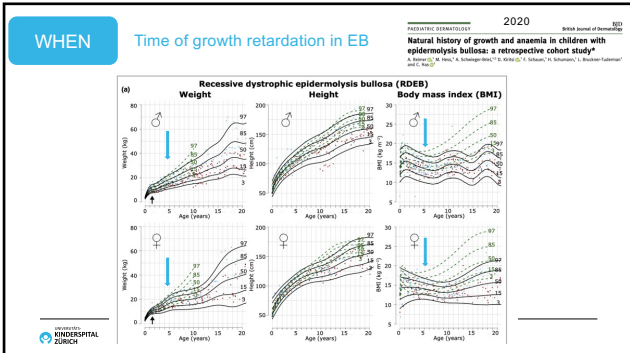
Main reasons for needing a G-tube in EB

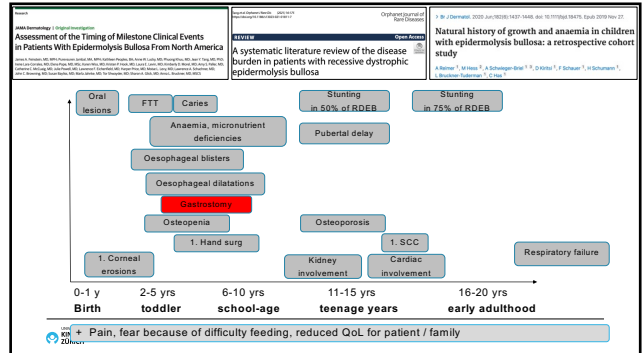




UNIVERSITÄT
KINDERSPITAL
ZÜRICH

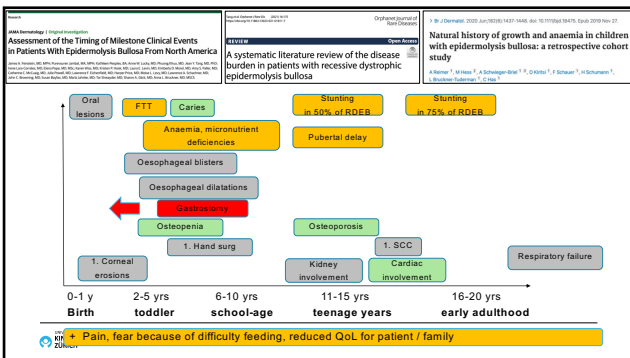
6



7



8



9

WHAT ELSE TO TAKE INTO ACCOUNT / TO ADDRESS ?

QoL in EB (affected individuals, caregivers)

- G-tube leads to improved QoL and high satisfaction in caregivers and patients
- despite complications
- despite inconsistent weight gain and nutritional advantages
- Less stress and more pleasure during meal times

• Consider risk of recurrence of esophageal strictures (>1 strictures, >1cm!)

Side Effects of G-tubes:

- Local irritations:** infections, eczema, leakage, pain
- GI tract:** Gagging, vomiting, dumping syndrome
- Tube issues:** discomfort, dislocation, blockage, perforations
- Others:** Feeding disorders, selectivity, tube dependency, non-acceptance of tube

• **Social consequences:** worries of issues with child care, visibility, stigmatization

Krom et al Eur J Pediatr (2017) Zidonio et al- BJD (2018) ; Colomb et al. BJD (2012) Kleinman et al. PedDerm (2022) Pope et al. Gastroenterology 2020; Haynes et al. Pediatr Dermatol (2012)

10

THE EARLIER THE BETTER?

- FTT predictable, especially in RDEB gen sev
- Optimal, balanced feeds early on
- Acceptance of G-tube likely better if performed early
- NG tube feeds in phases of blistering can lead to trauma and food aversion
- Episodes of blistering are scary for children and caregiver → G-tube offers autonomy

- Significant development of feeding skills in 1st year of life → interrupted with early G-tube feeding → increased risk of tube dependency
- Reduced sensory stimulation and impaired social interaction at meal times
- Oral disorders in 7/9 pts after early G-Tube insertion in EBS (3,7 mo)
- Parental acceptance requires certain experiences, takes time!

Krom et al Eur J Pediatr (2017); Marro et al. Orphanet J Rare Dis (2021) Bagela et al. Clin Exp Dermatol (2024); Riccio; Clin Nutrition (2015)

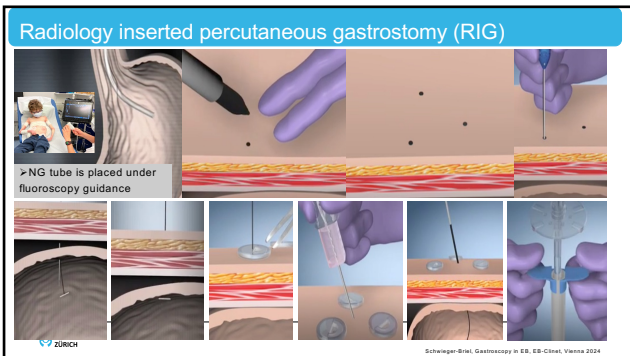
11

HOW – Different ways of gastrostomy insertion in EB

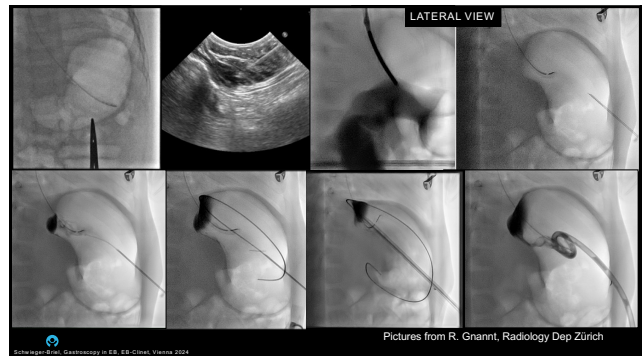
- Surgical insertion → direct → rarely performed
- Percutaneous endoscopic gastrostomy (PEG)
- Radiology inserted / Primary image guided gastrostomy (RIG)

Schwager-Briel, Gastroscopy in EB, EB-Clinical, Vienna 2024

12



14



15

Advantages of RIG

- > Does not require deep general anaesthesia
- > Low complication rates (similar as PEG)
- > Less esophageal shearing forces → less damage during procedure
- > Can be done with strictures (only fine gastric tube needs to pass)
- > Fast and cheap

Disadvantage at this point: **2 step procedure**

→ change to a button after 2 months

KINDERSPITAL ZÜRICH

16

Future

Journal of Medical Imaging and Radiation Oncology 67 (2023) 519-525

INTERVENTIONAL RADIOLOGY—ORIGINAL ARTICLE

An 8-year, single-centre experience of primary image-guided insertion of 'button' gastrostomy catheters: Technical and clinical results

Mark Sheehan,^{1,2} Cormac O'Brien,^{1,2} Desmond Killick,¹ Hayley Briody,¹ Aoife Keeling,¹ Mark Given,¹ Andrew McGrath¹ and Michael J Lee^{1,2}

¹ Department of Radiology, Beaumont Hospital, Dublin, Ireland
² Royal College of Surgeons in Ireland, Dublin, Ireland

KINDERSPITAL ZÜRICH

17

2 cases

KINDERSPITAL ZÜRICH

18


Louis, 2 yrs old

- RDEB intermediate generalized
- Difficulty feeding
- Episode of acute esophageal blistering at age 9 mo → NG tube
- Parents ask for G-tube

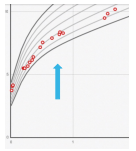
KINDERSPITAL ZÜRICH

19

Louis *2022



- Only 9 months old
- Preexisting difficulty feeding → possibly not associated with RDEB
- No FTT
- No stricture
- → no G-tube at this point



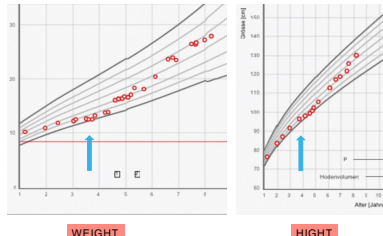
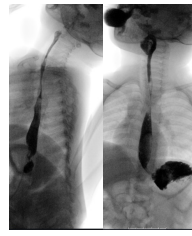
WEIGHT

KINDERSPITAL ZÜRICH

20

Jonas *2016

- RDEB severe generalized, mild-moderate
- 1st dilatation at age 3,5 yrs
- FTT

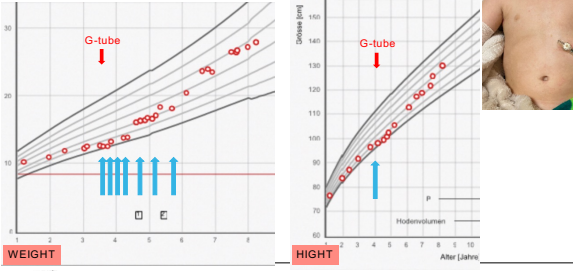




WEIGHT **HIGHT**

KINDERSPITAL ZÜRICH

23

Eats for pleasure (1/3 of his needs) plus daily boluses + nightly pump feeds





WEIGHT **HIGHT**

KINDERSPITAL ZÜRICH

24

G-tube in EB



Decision for a G-tube has to be made on an individual basis

Depending on

- **symptoms** (e.g. pain, FTT, constipation etc)
- **type of EB** and associated expected complications
- **length and number of esophageal strictures** → frequency of dilatations
- **stress level in the family**, family /patient wishes

→ Best age seems to be in **early childhood** (appr. 2-3 years of age)

→ Ideally **AFTER** normale food intake, eating habits have been learned

→ Families need to be **„ready,“ well informed** and have **realistic expectations**

KINDERSPITAL ZÜRICH

25

Thank you for your attention



EB-KidZ Team Zürich
ebteam@kispi.uzh.ch

KINDERSPITAL ZÜRICH Schwagerl-Diet, Gastroscopy in EB, EB-Cinet, Vienna 2024

26