International Intestinal Failure Registry 2025 Report

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IIFR
International Intestinal Failure Registry

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NASPGHAN

aspen LEADING THE SCIENCE AND PRACTICE OF CLINICAL NUTRITION
American Society for Parenteral and Enteral Nutrition

Disclosures

- Takeda LTD
- Zealand Pharma
- Ipsen innovation SAS
- SterileCare Inc.

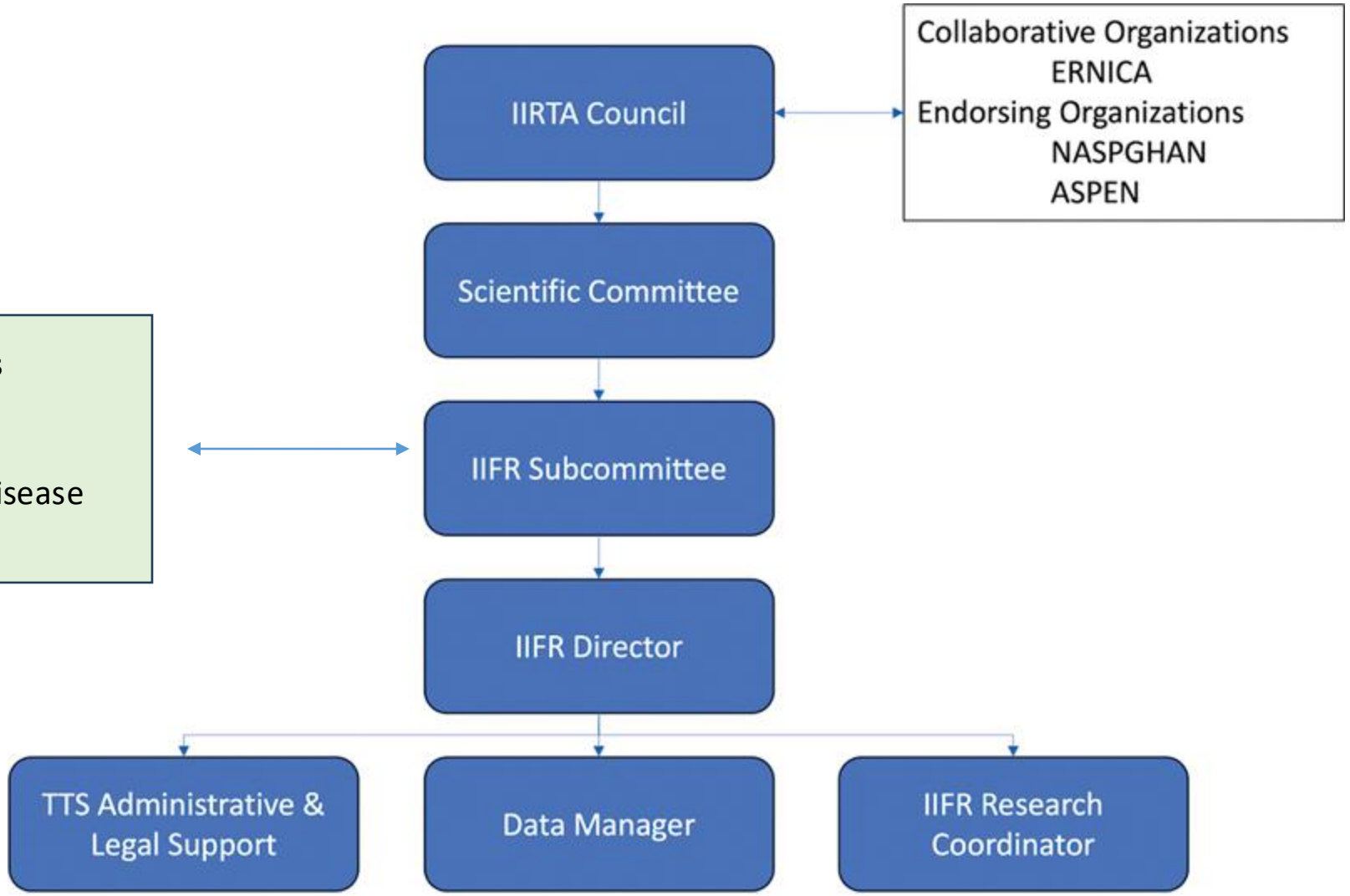
Goals

Creation of a large, international data base of pediatric intestinal failure to inform on worldwide trends and outcomes beyond single center experiences

Creation of benchmarking and learning networks in pediatric intestinal failure

Identification of favorable interventions and treatments to inform on best practices and evidence-based treatment approaches

IIFR Structure



Research networks
- Local
- International
- FDA RO1 rare disease grant

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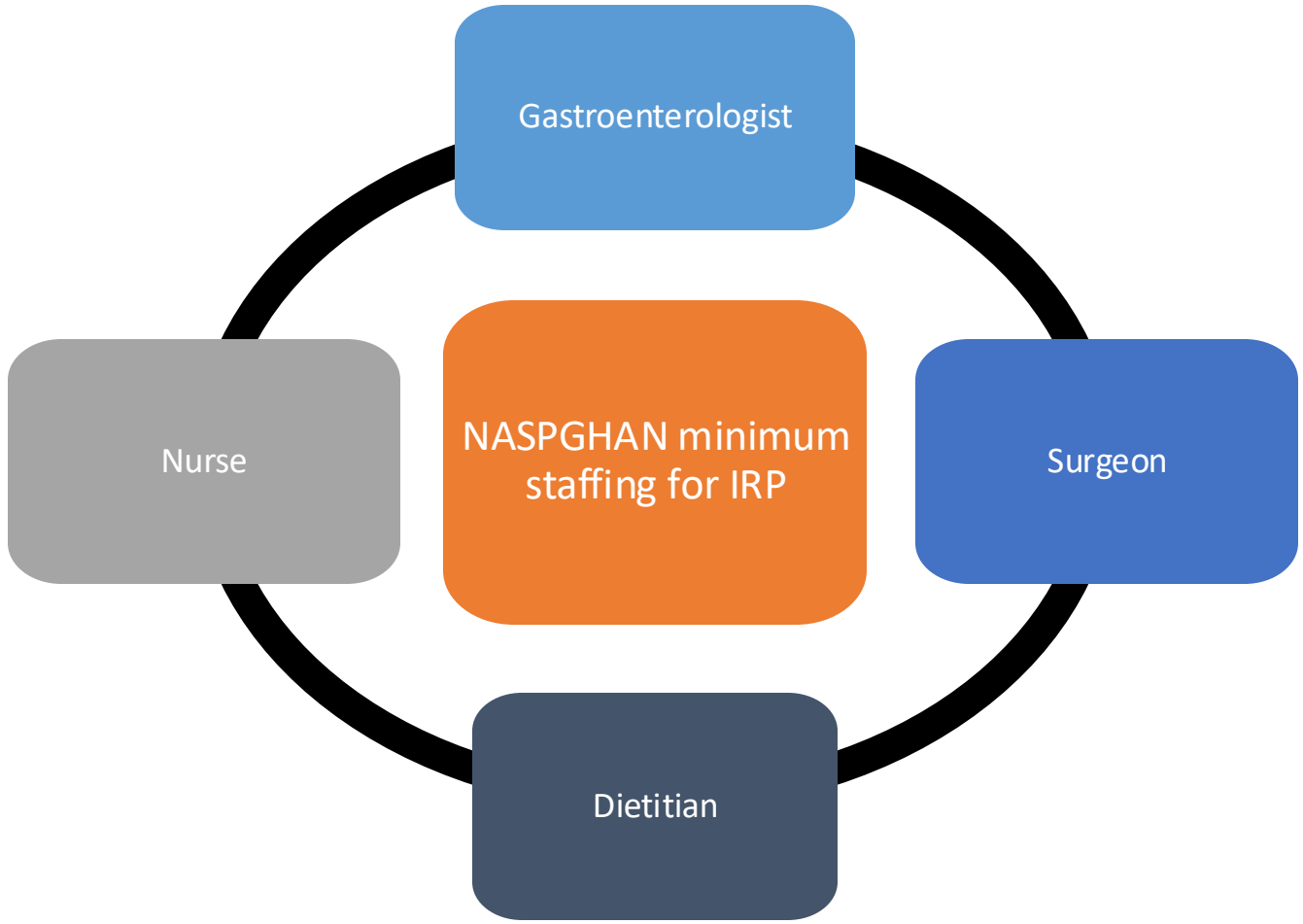
Avitzur et al. EJPS 2024

IIFR - Structure

- Prospective data collection
- REDCap based data collection
- Open to all centers with a pediatric intestinal rehabilitation program as defined by NASPGHAN position statement (Minimum of a Gastroenterologist, surgeon, dietitian and a nurse)*



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Merritt RJ, et al. *JPGN*. 2017;588

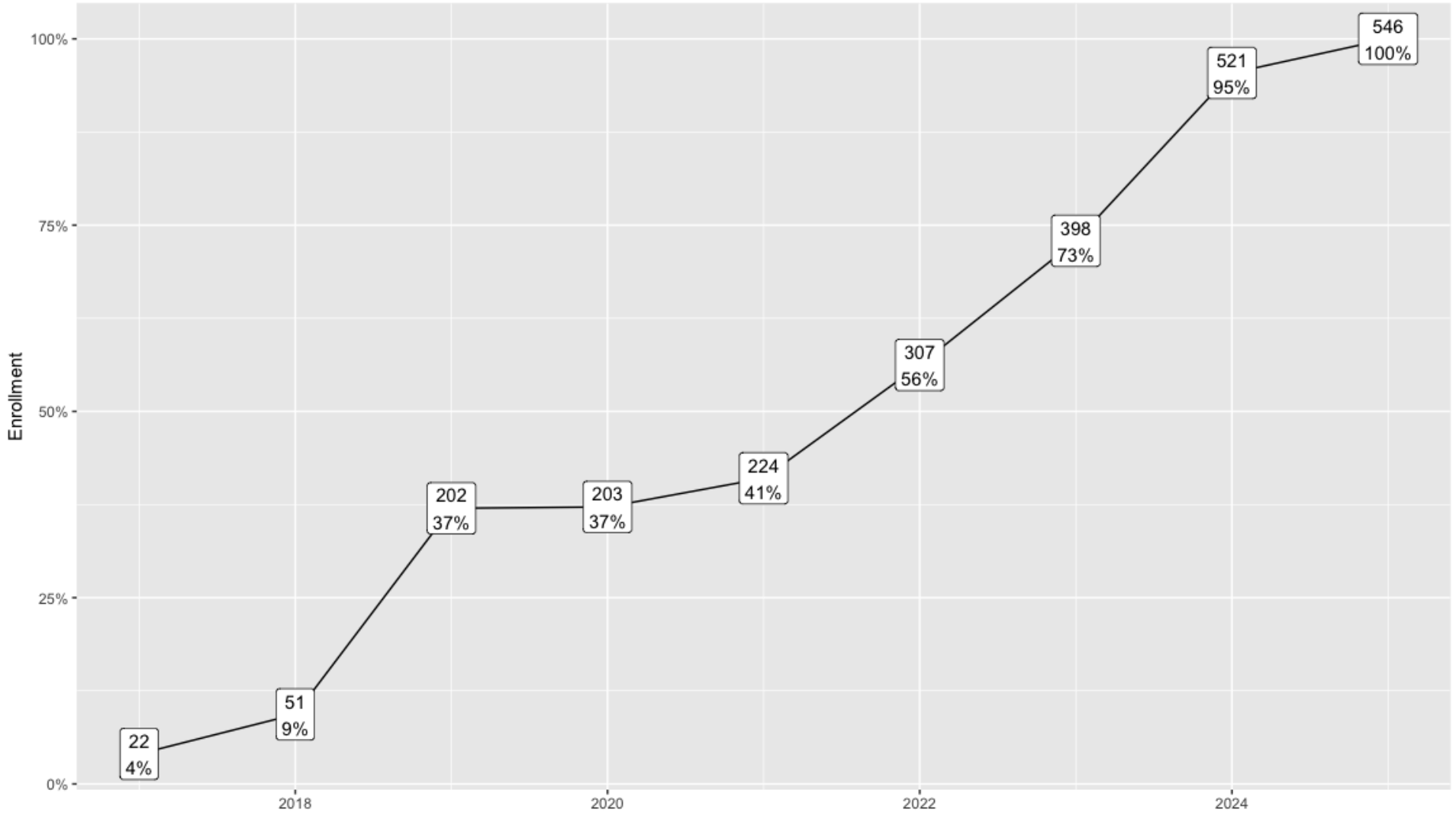
IIFR - Eligibility

Inclusion criteria

- Children younger than 18y with intestinal failure (≥ 60 days on parenteral nutrition defined as intravenous solution containing protein, carbohydrate, electrolytes and trace elements / vitamins)
- Primary underlying gastrointestinal disorder. IF etiologies can include short bowel syndrome, motility disorders and congenital diarrheas and enteropathies
- Followed by intestinal rehabilitation program as defined by NASPGHAN position statement 2017(minimal program staffing of a pediatric gastroenterologist, surgeon, dietitian and a nurse)

Minimal annual patient volume is not required

Annual Patient Enrollment



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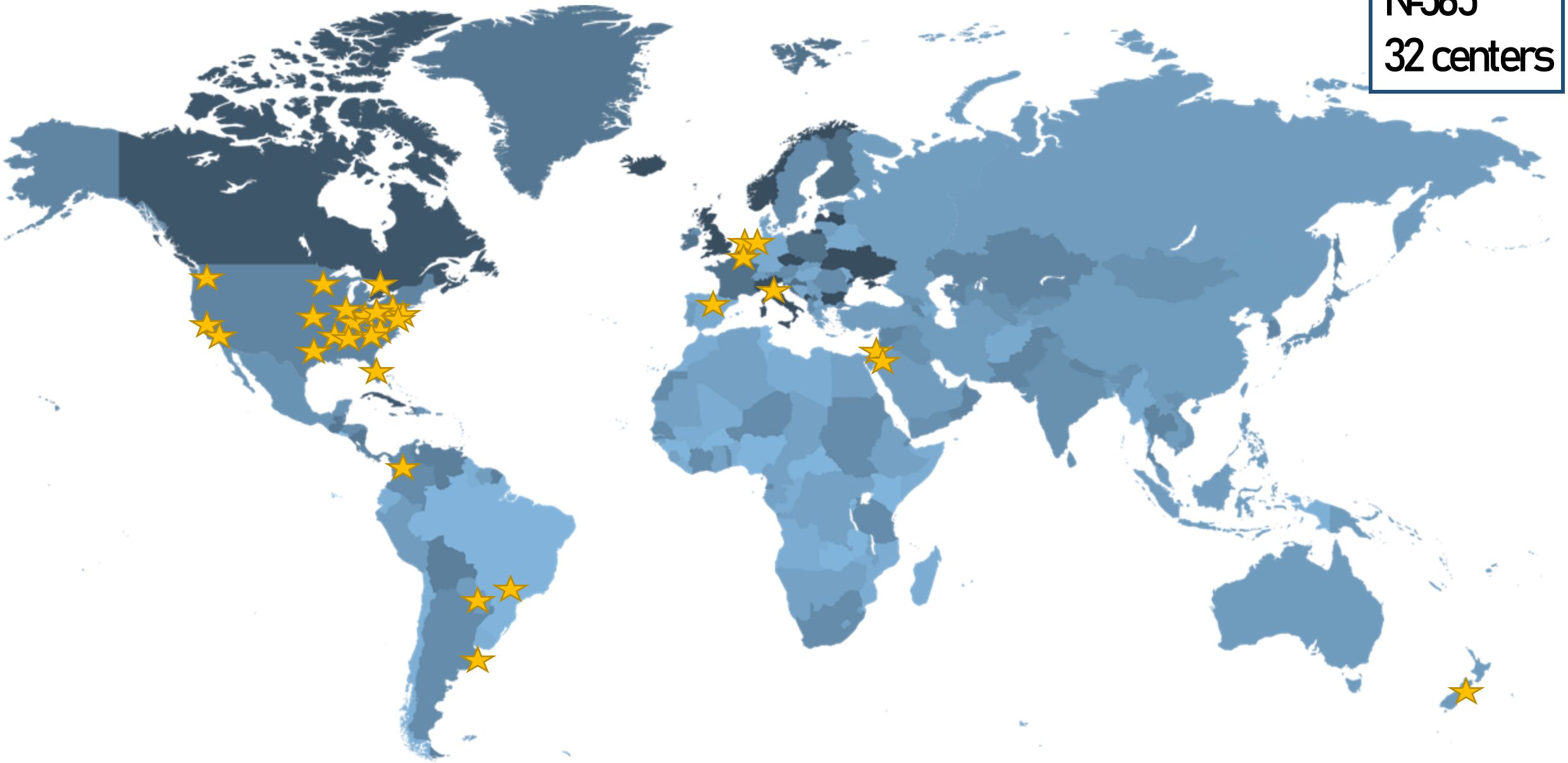

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Annual IIFR Center Report

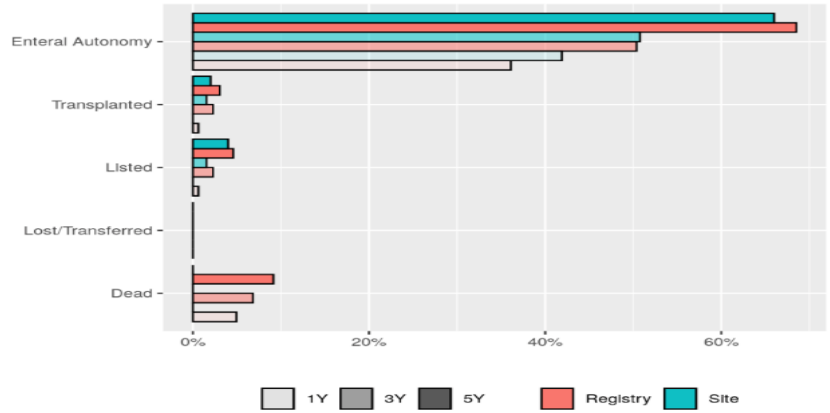
Center specific report and IIFR average for the same data points

- Number of patients recruited – total and annual
- **Quality parameters**: Field completion rate (<80% & <50%); number of missing visits
- **Patient demographics**: Age & IF etiology
- **Overall outcome**: Enteral autonomy; Transplant and death; Overall and at 1, 3, 5y since diagnosis
- **Functional outcomes**: Lansky score & % of oral aversion
- **Specific outcomes**: rate of IFALD, vascular thrombosis, CLABSI /1000PN days

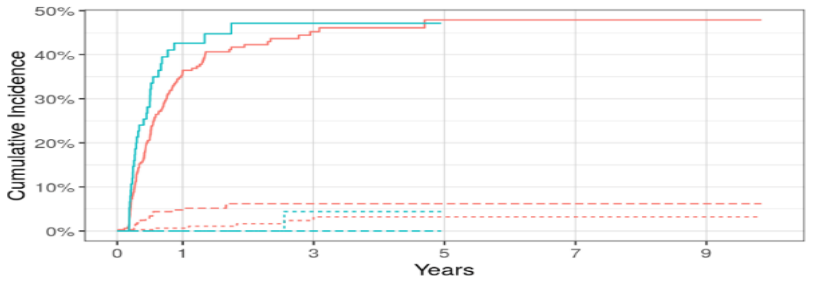
Annual Center Report

International Intestinal Failure Registry Annual Center Report | Sample | May 2023 | Cases | Outcomes | Quality

Outcomes 1Y, 3Y, 5Y from enrollment



Cumulative Incidence



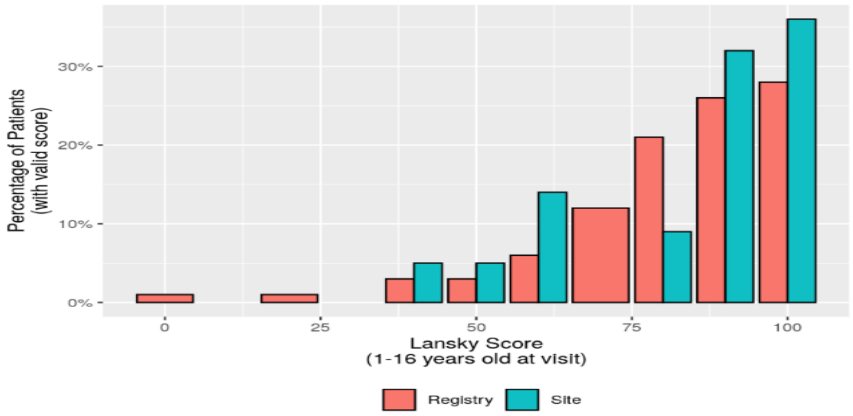
Registry

At Risk	337	142	55	22	14	11
Events	0	130	152	154	154	154

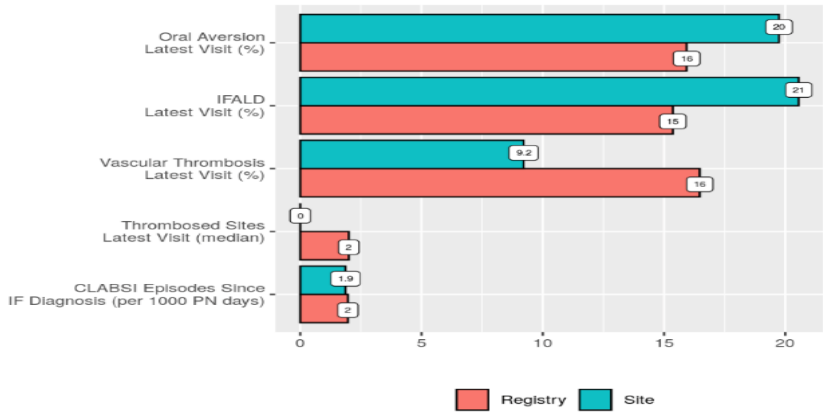
Site

At Risk	76	34	9	0	0	0
Events	0	31	34	34	34	34

Functional Outcomes



IF Complications



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IIFR – Publications (2023-2024)

DOI: 10.1002/jpen.2557

ORIGINAL RESEARCH

Predictors of 1-year enteral autonomy in children with intestinal failure: A descriptive retrospective cohort study

Vikram K. Raghu MD, MS[1] | Harold J. Leraas MD, MHS[2] | Mariya Samoylova MD, MAS[2] | Christine Park BS[2] | Scott D. Rothenberger PhD[3] | Debra Sudan MD[2] | Yaron Avitzur MD[4]

JPEN 2023;1047-1055

The Development of the International Intestinal Failure Registry and an Overview of its Results

Yaron Avitzur[1] Eric Pahl[2] Robert Venick[3,4] and the International Intestinal Failure Registry

Eur J Pediatr Surg 2024;172-181

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IIFR-Ongoing Research Projects

First results from the IIFR: Therapeutic management and outcomes of chronic intestinal failure in Europe

PI: Dr. Tabbers, Emma Children's Hospital, Amsterdam, The Netherlands

Prevalence of education needs for children with intestinal failure

PI: Dr. Alaish, Johns Hopkins, Baltimore, MD, USA

Racial and ethnic disparities in pediatric intestinal failure

PI: Dr. Wadhwani & Gutierrez, UCSF, San Francisco, CA, USA

Disease course and risk stratification of IFALD in pediatric patients: A longitudinal registry-based analysis

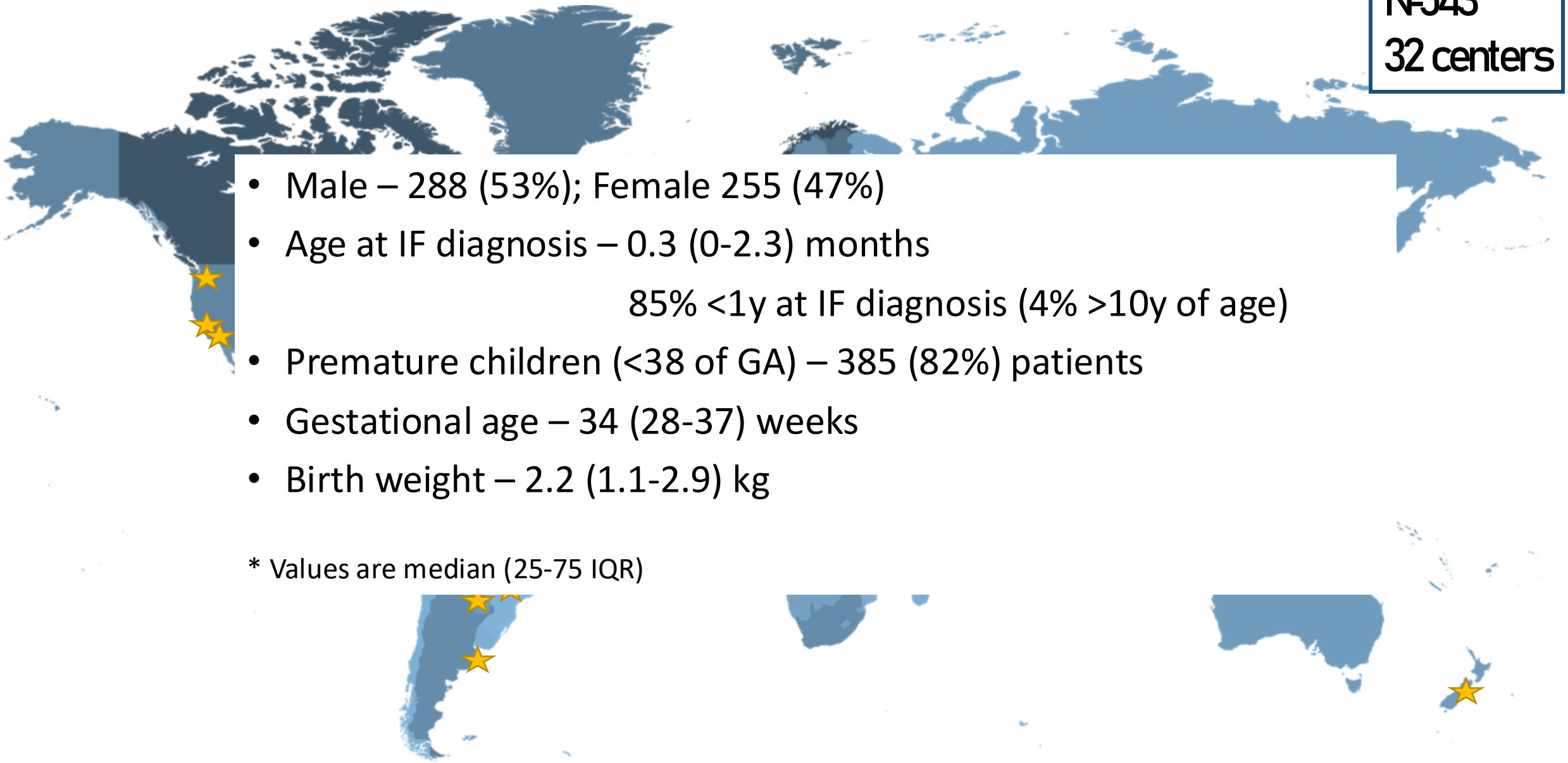
PI: Dr. Avitzur, SickKids, Toronto, Canada

IIFR- FDA RO1 Rare Disease Grant

Aim 1: Describe the natural history of IF in the current era of IF management

Aim 2: Assess PROMs using a novel IF community-derived disease specific tool

Aim 3: Develop disease specific outcome measures through machine learning techniques



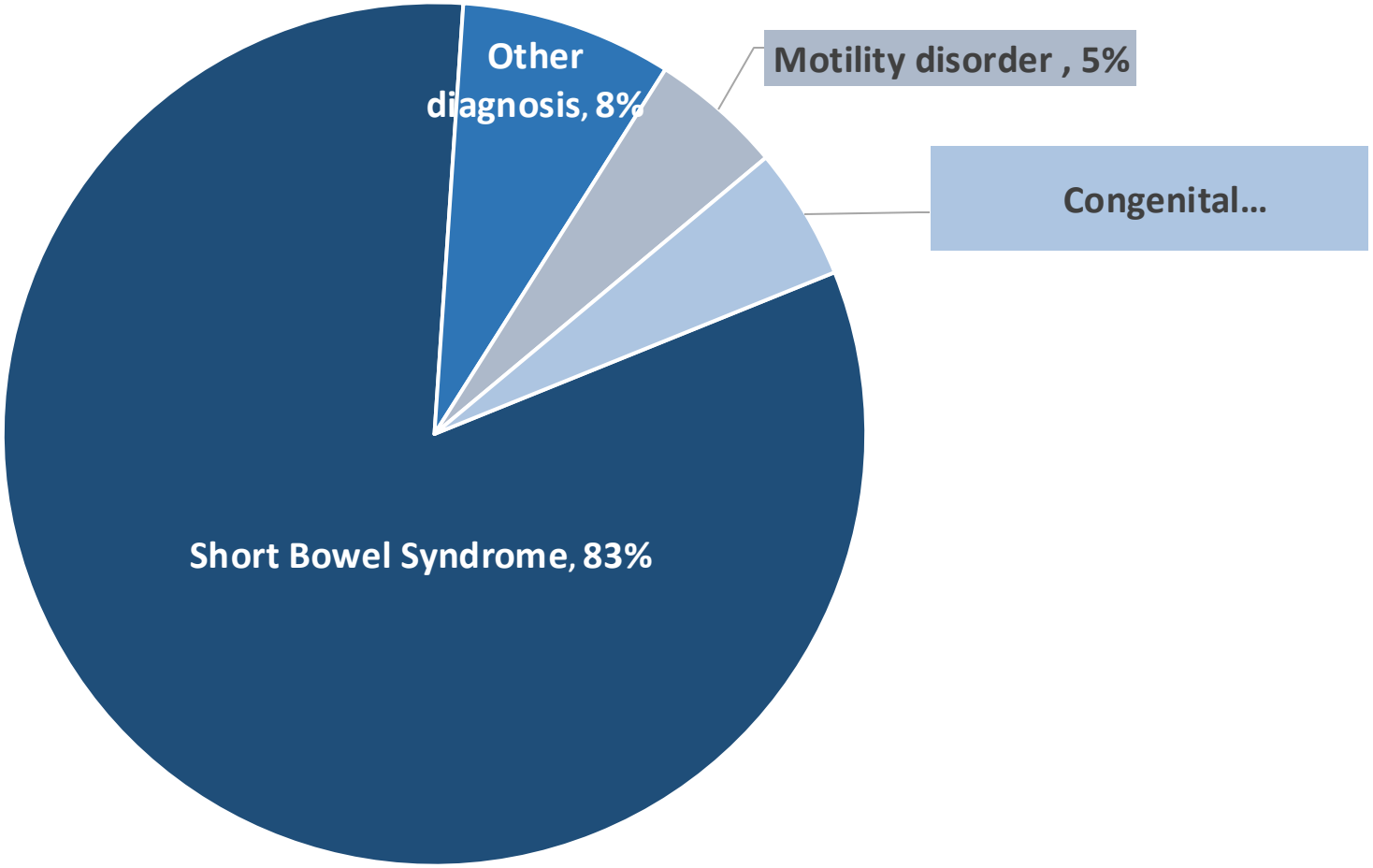
N=543
32 centers

- Male – 288 (53%); Female 255 (47%)
- Age at IF diagnosis – 0.3 (0-2.3) months

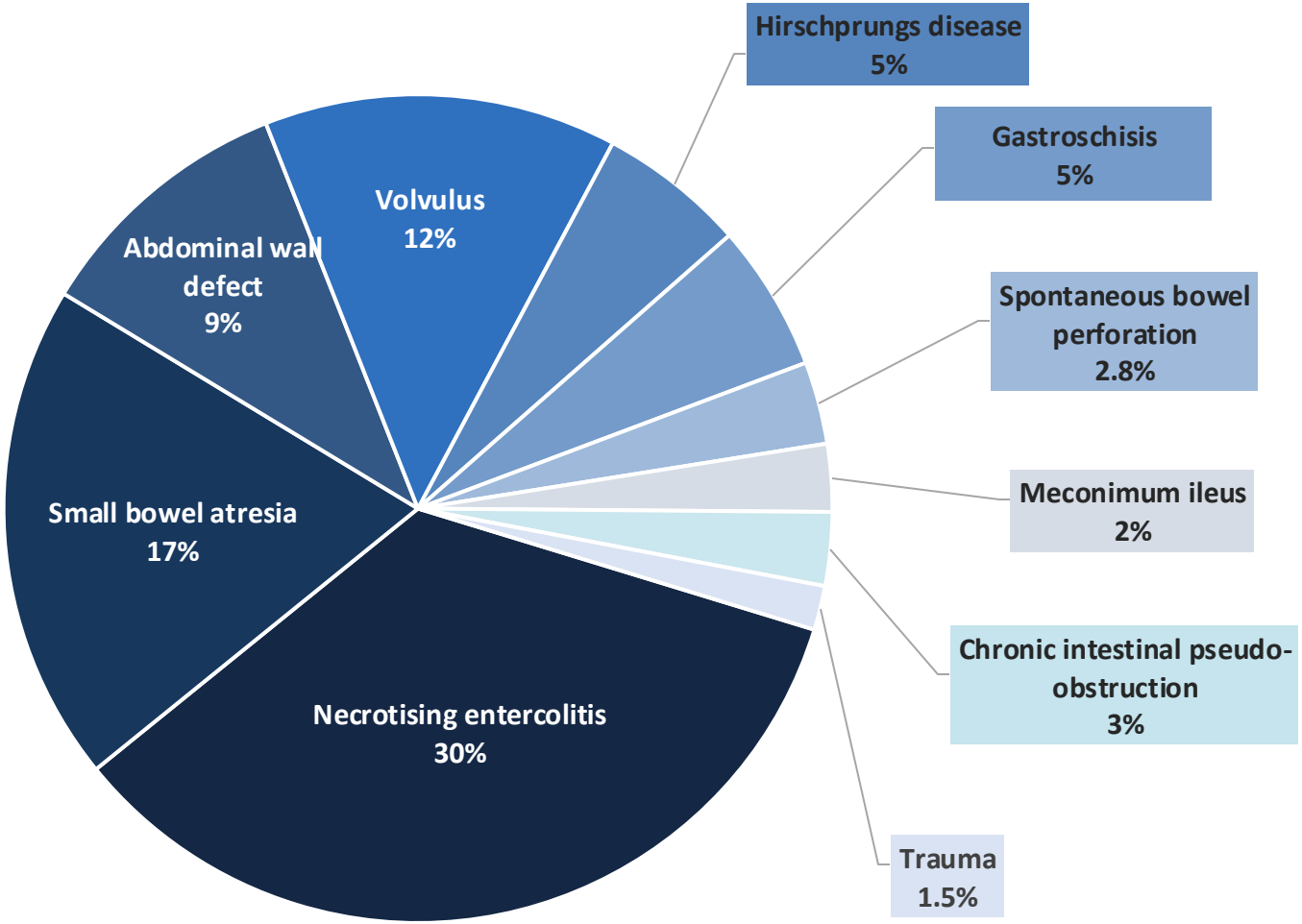
  85% <1y at IF diagnosis (4% >10y of age)
- Premature children (<38 of GA) – 385 (82%) patients
- Gestational age – 34 (28-37) weeks
- Birth weight – 2.2 (1.1-2.9) kg

* Values are median (25-75 IQR)

Primary Intestinal Failure Etiology

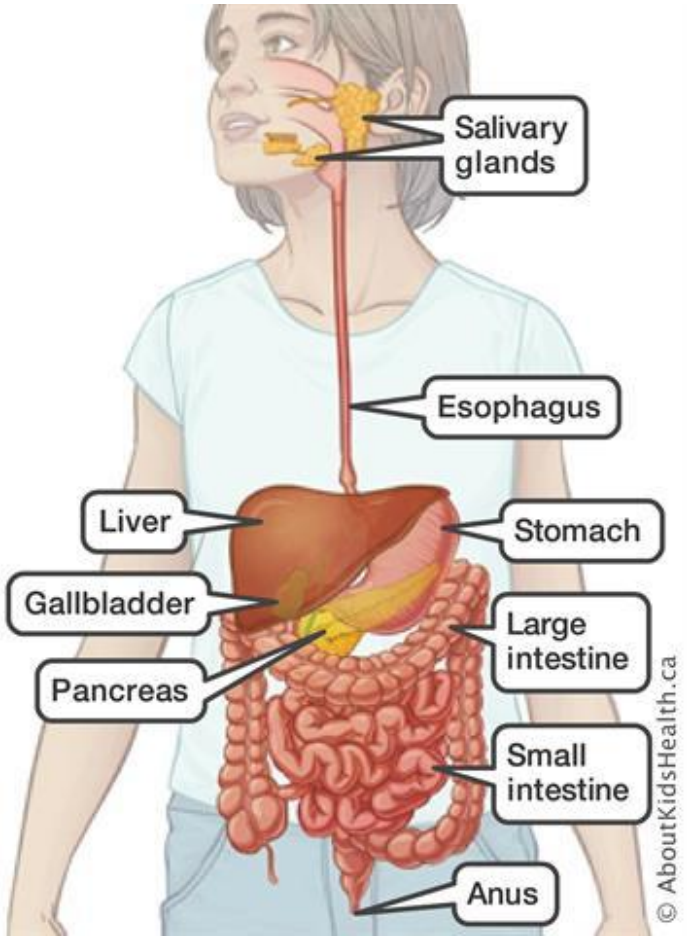


Primary SBS Etiology



Bowel Anatomy

- Patients that have had bowel resection at baseline or any follow-up: n = 445 (82%)
- Shortest small bowel length at baseline or any follow-up: 40 (20-65) cm (N=327)
- Percentage of expected small bowel length per age: 19% (10% - 41%)
- Ileocecal valve resection at baseline or any follow-up: n = 203 (37%)
- 71% with intact colon at 2m after diagnosis
- Patients with stoma at baseline or any follow-up: n = 261 (48%)



* Values are medians (25-75 IQR)

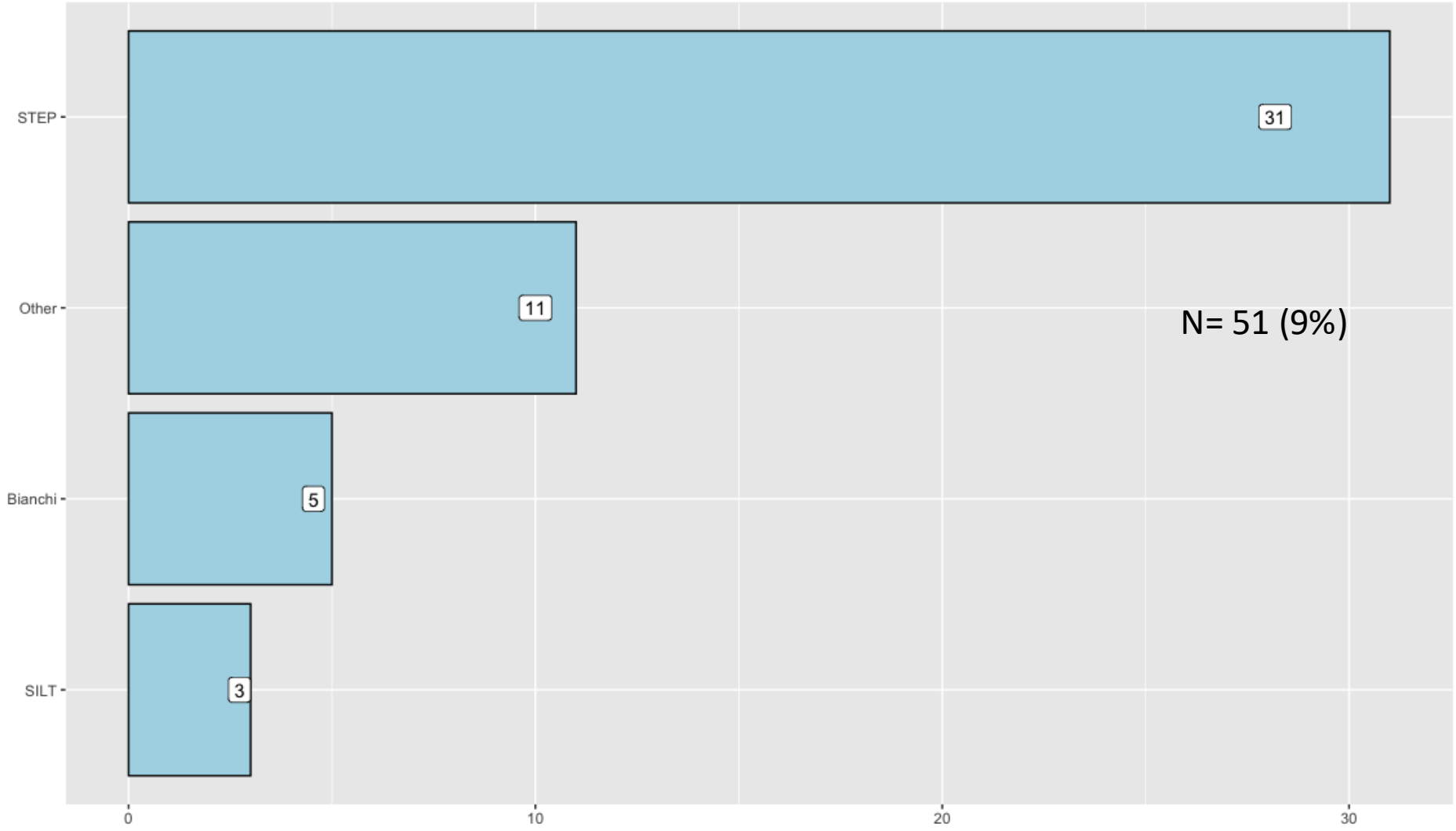
https://www.aboutkidshealth.ca/Article?contentid=4061&language=English

Surgeries Post Initial Resection

visit	n	Bowel resection	Creation of stoma	Reversal of stoma	Bowel plication	Bowel lengthening	Other surgery
6 month	467	57 (12%)	32 (7%)	95 (20%)	2 (0%)	7 (1%)	50 (11%)
1 year	346	7 (2%)	5 (1%)	18 (5%)	0	3 (1%)	32 (9%)
2 year	177	1 (1%)	1 (1%)	7 (4%)	0	3 (2%)	16 (9%)
3 year	96	4 (4%)	3 (3%)	2 (2%)	2 (2%)	3 (3%)	11 (11%)
4 year	58	0	0	0	0	0	2 (3%)
5 year	49	2 (4%)	0	1 (2%)	1 (2%)	1 (2%)	7 (14%)
Total	**467**	**71 (15%)**	**41 (9%)**	**123 (26%)**	**5 (1%)**	**17 (4%)**	**118 (25%)**

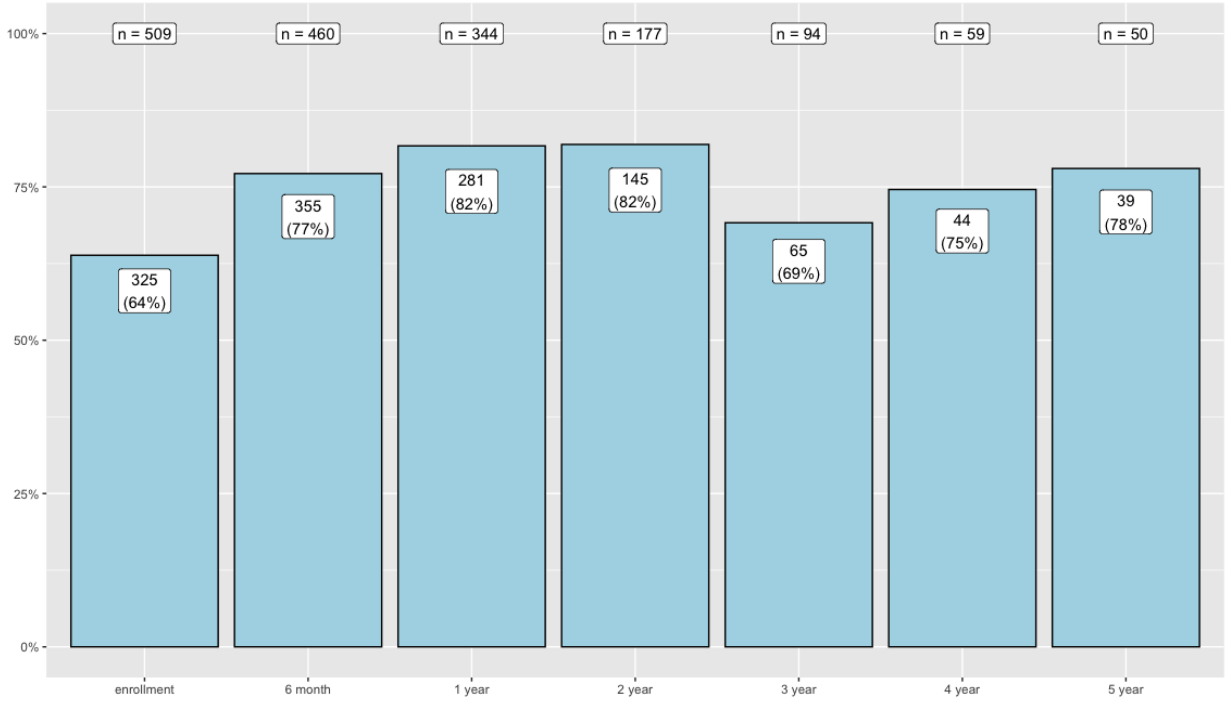
- Lengthening procedure at any time – 47 (9%)
- Median age at lengthening procedure – 16m (IQR 4-29m)

Lengthening Procedures – Type

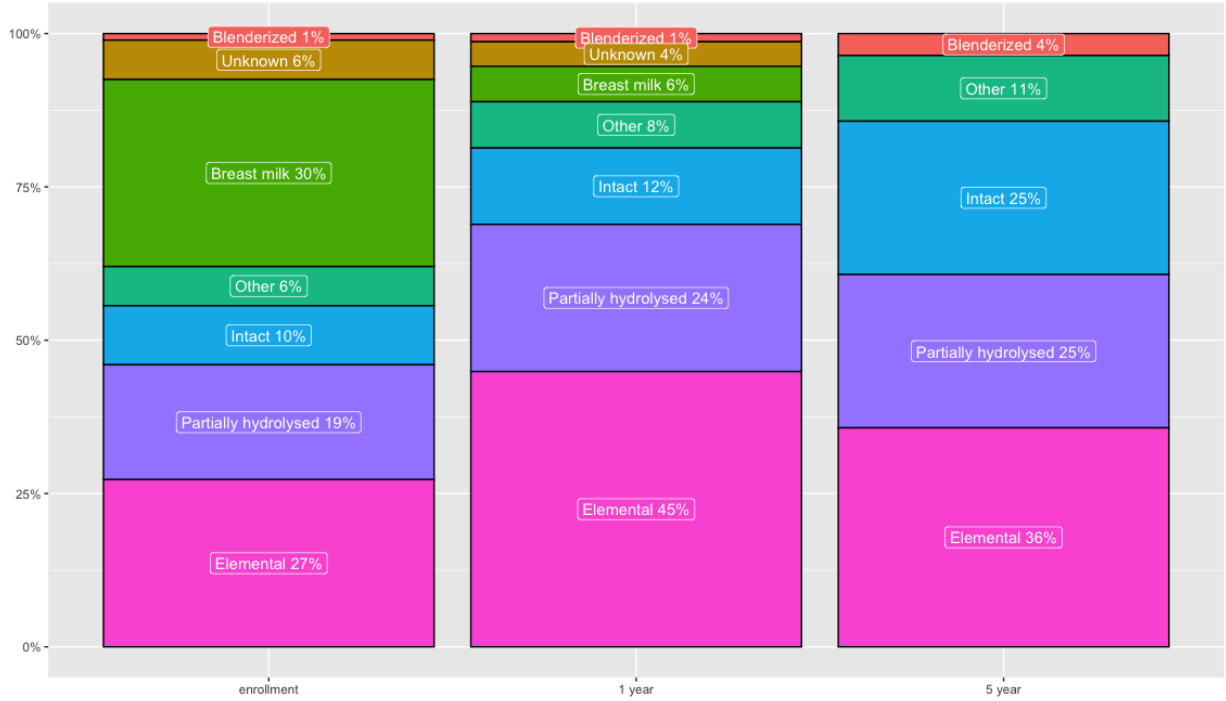


Enteral Nutrition

Use of enteral nutrition

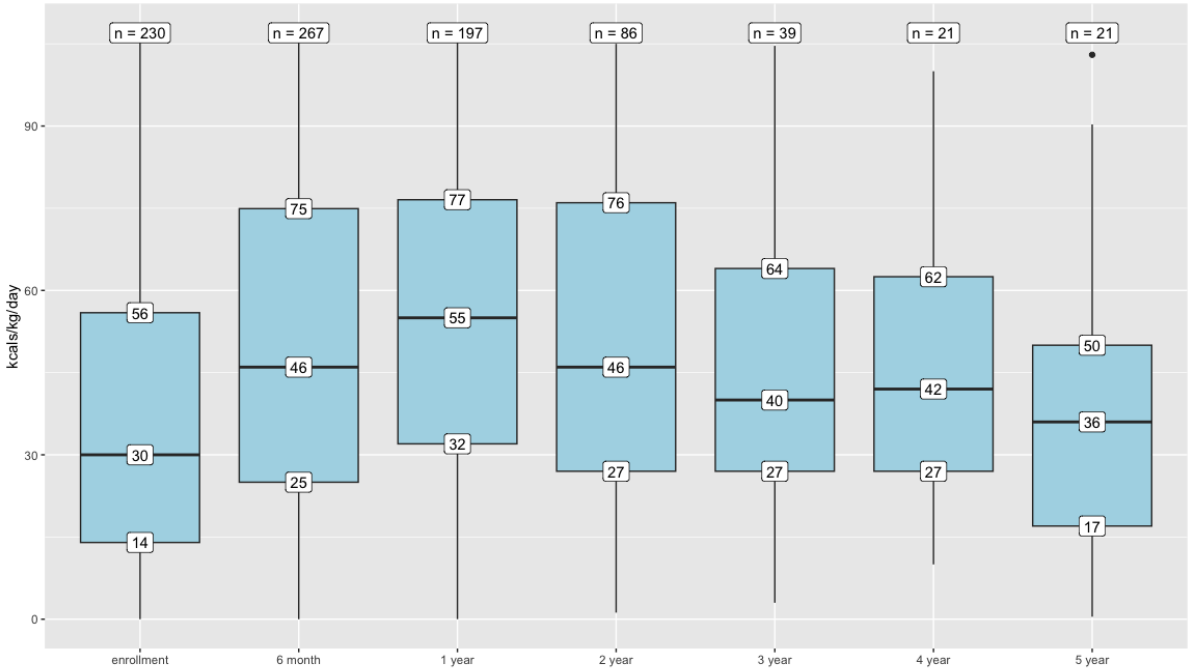


Type of formula

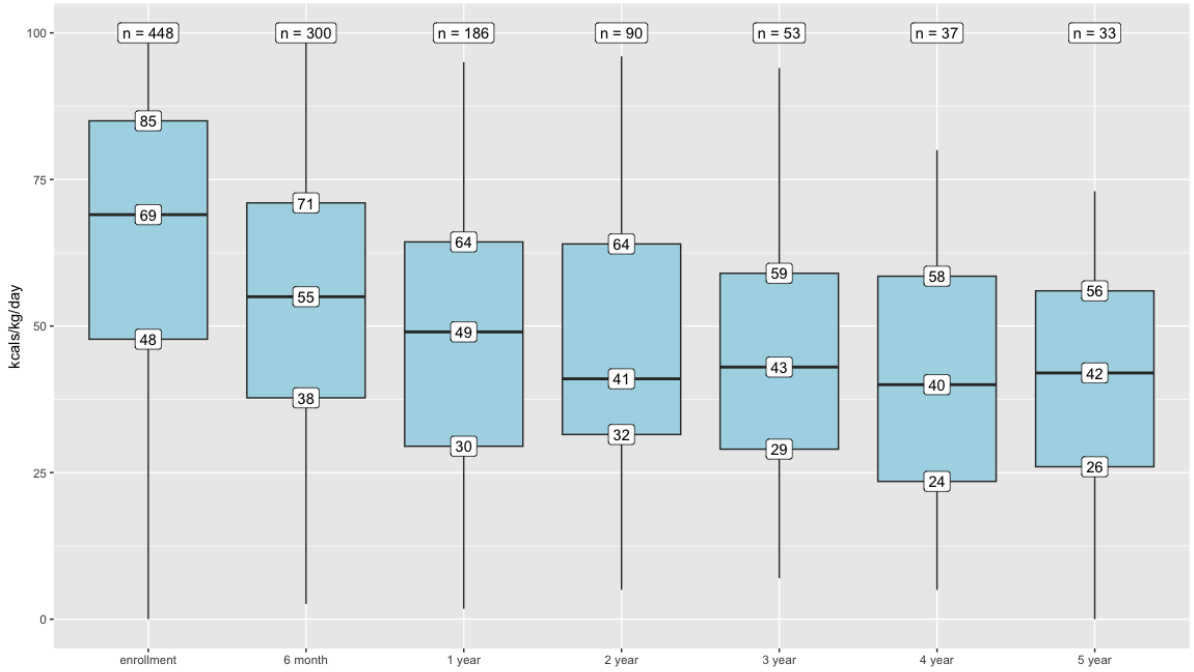


Nutrition – PN and EN Calories

Formula Calories (Kcal/kg/d)



TPN Calories (Kcal/kg/d)



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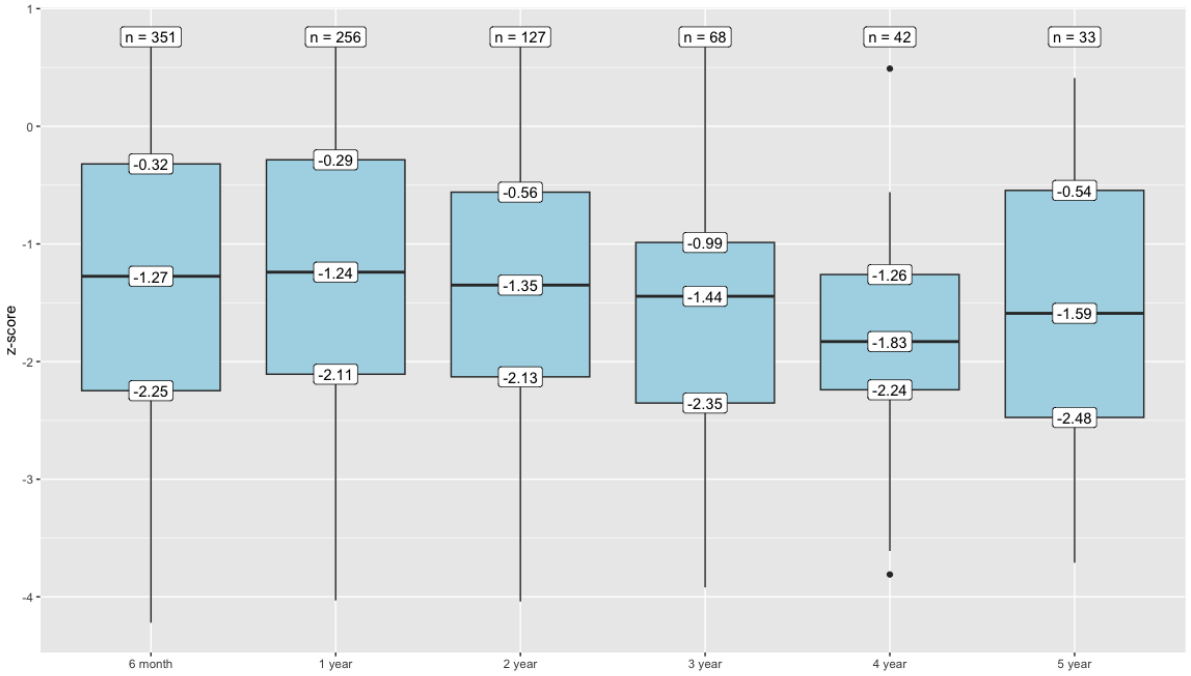
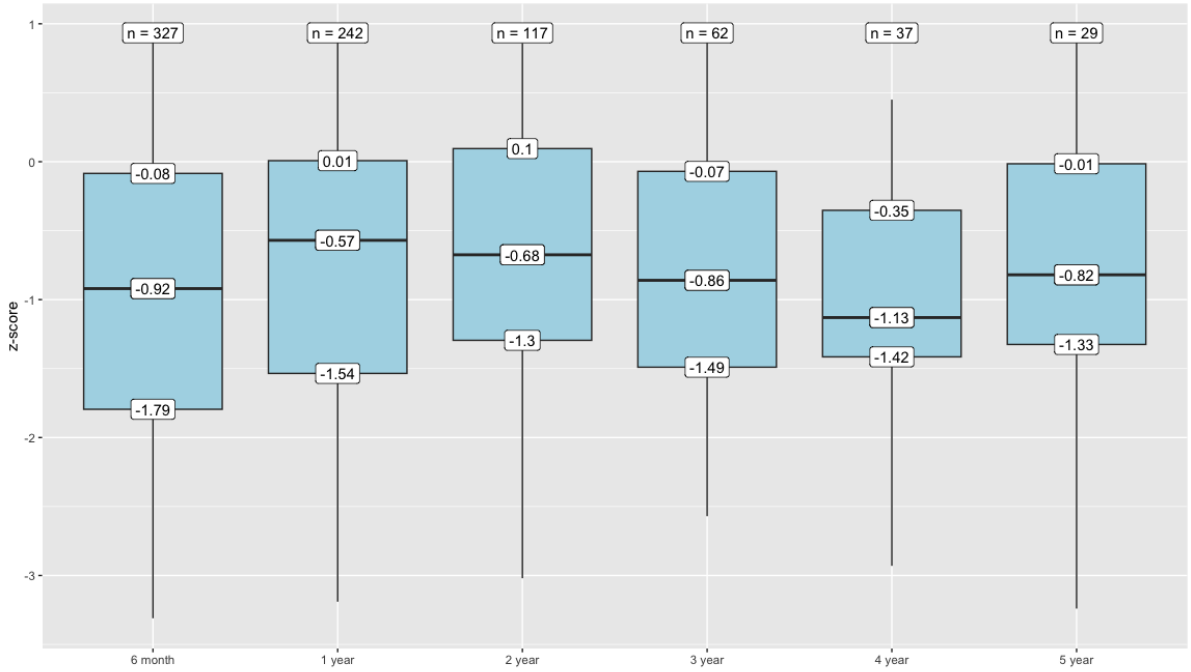
Growth

Height for age Z-score*



Weight for age Z-score*



* Adjusted age for premature infants <2y

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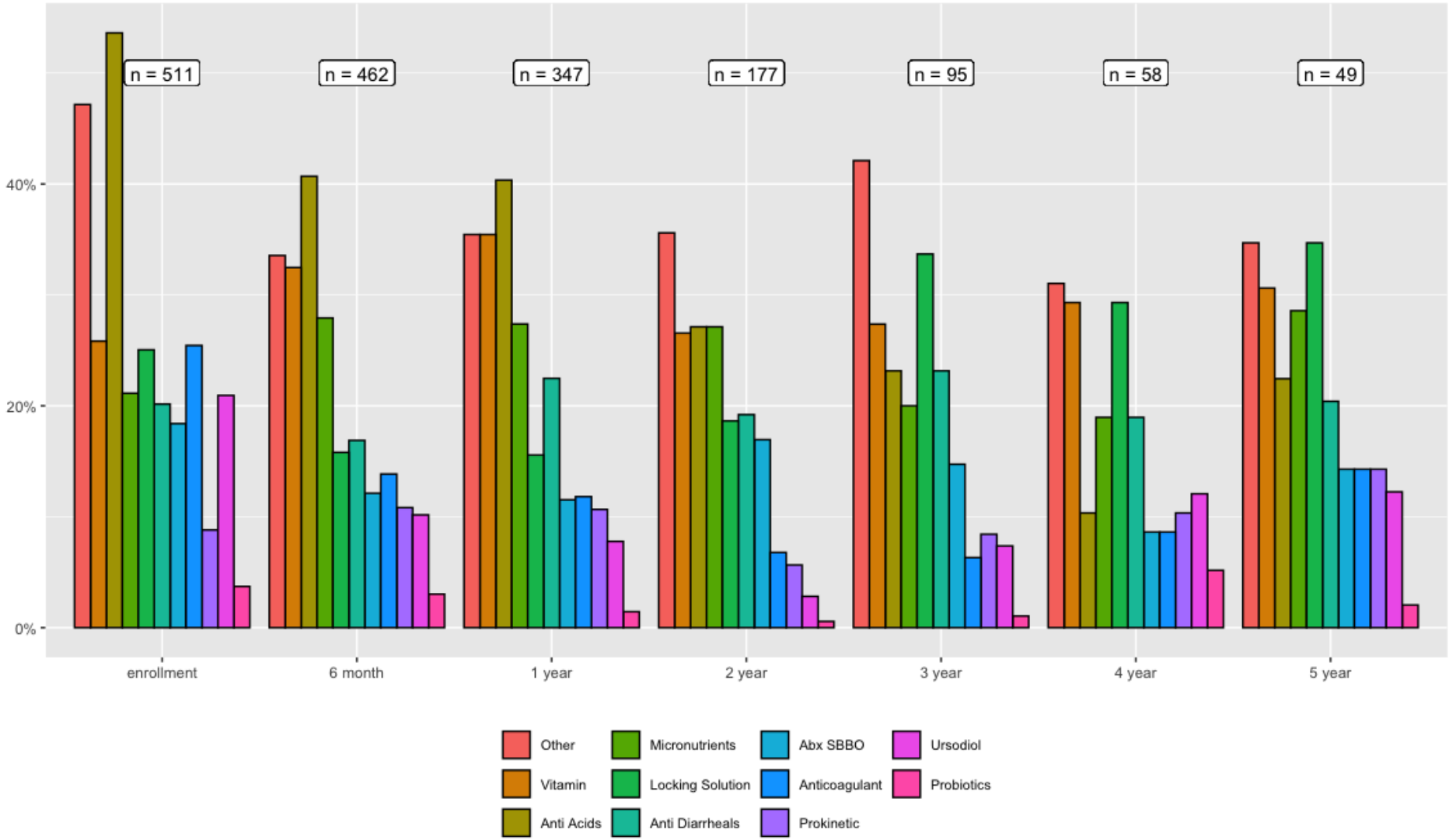
Medications



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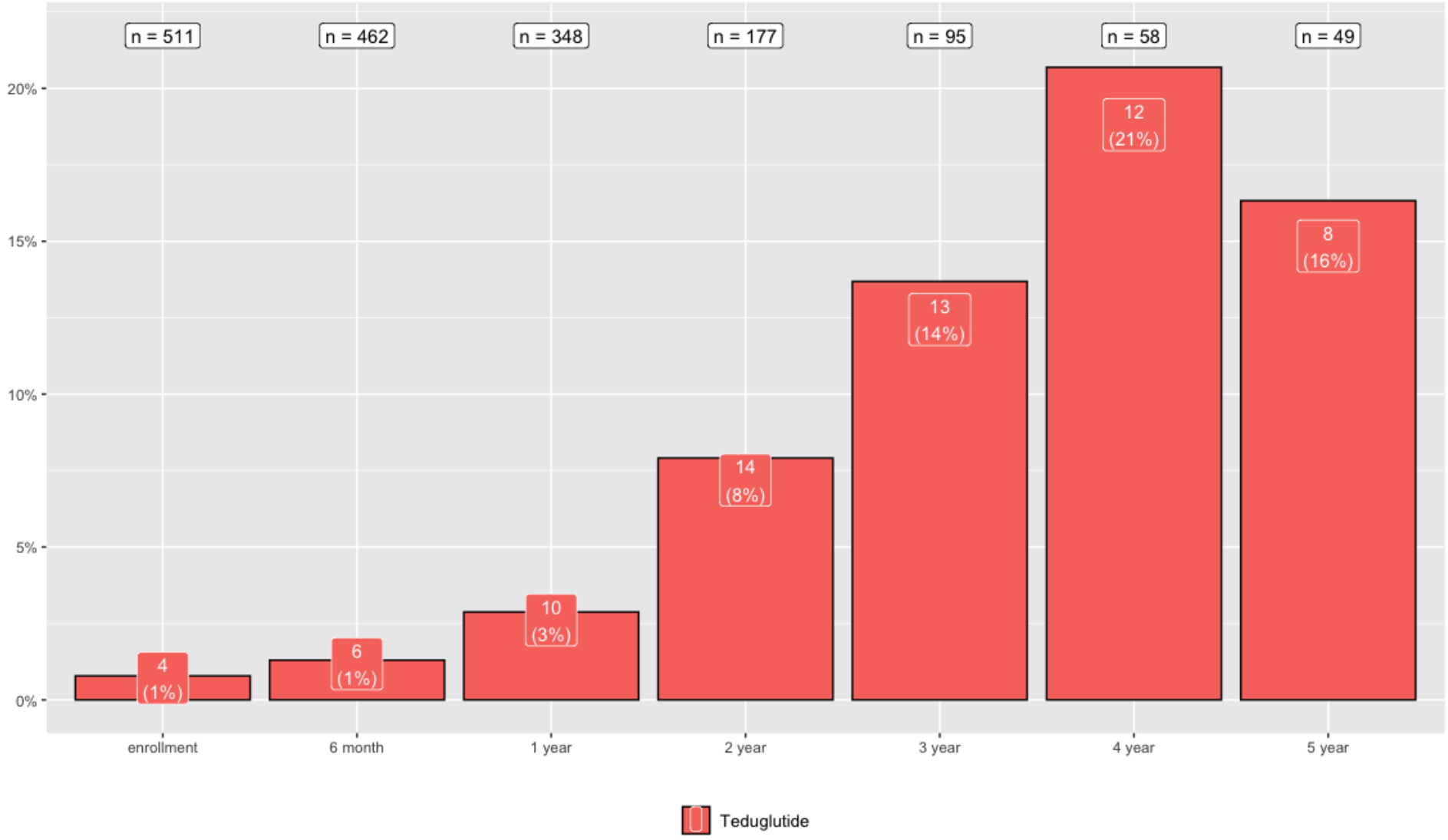
Teduglutide



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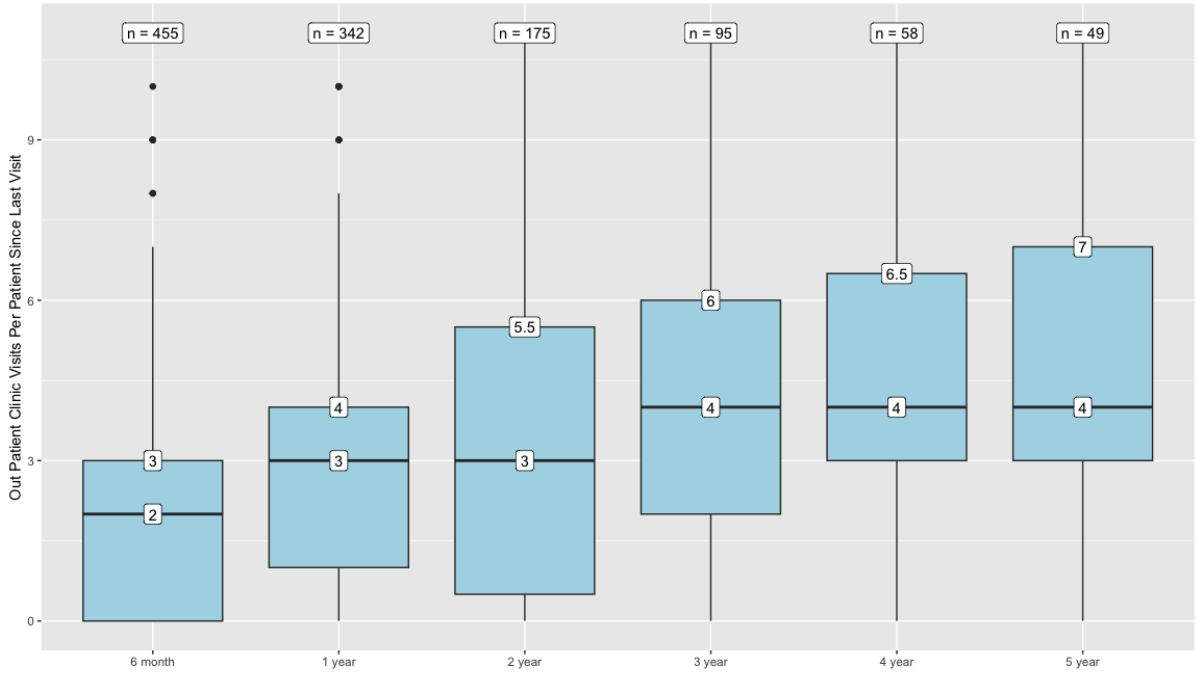
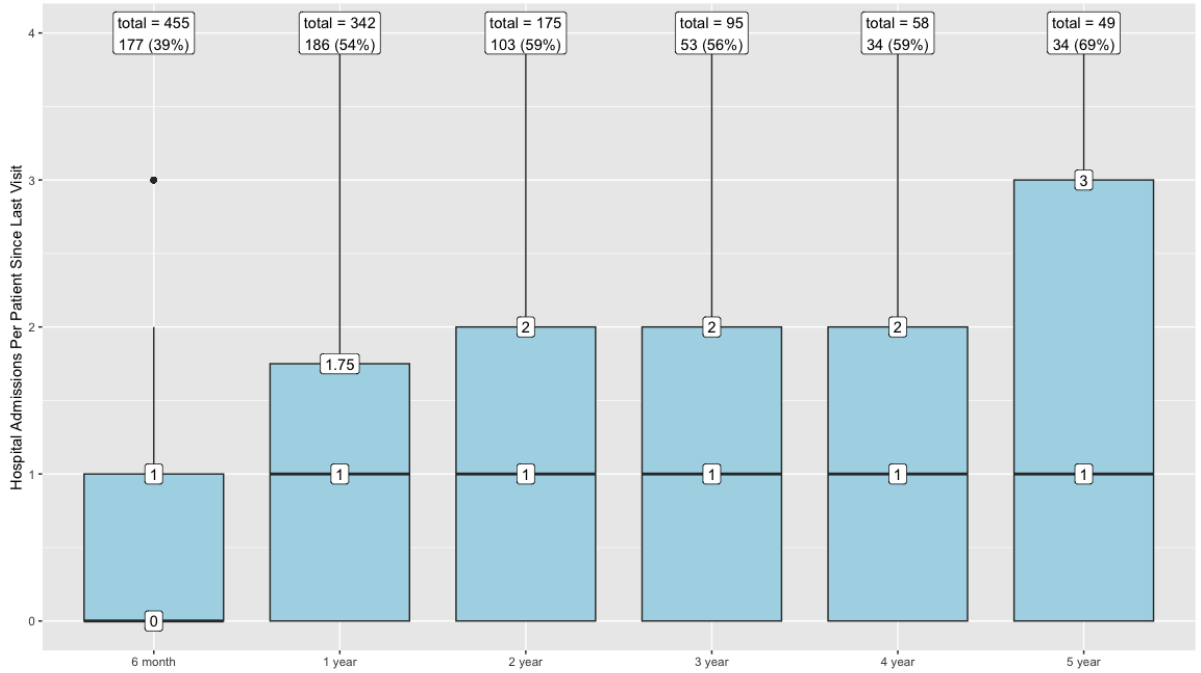
Health Care Utilization

- Discharge since first admission – 37% by 2m, 78% by 6m and 86% by the end of the first year
- Length of stay first admission – 3.9 (IQR 2.5-5.6) months

Clinic visits since last F/U



Hospital admissions since last F/U



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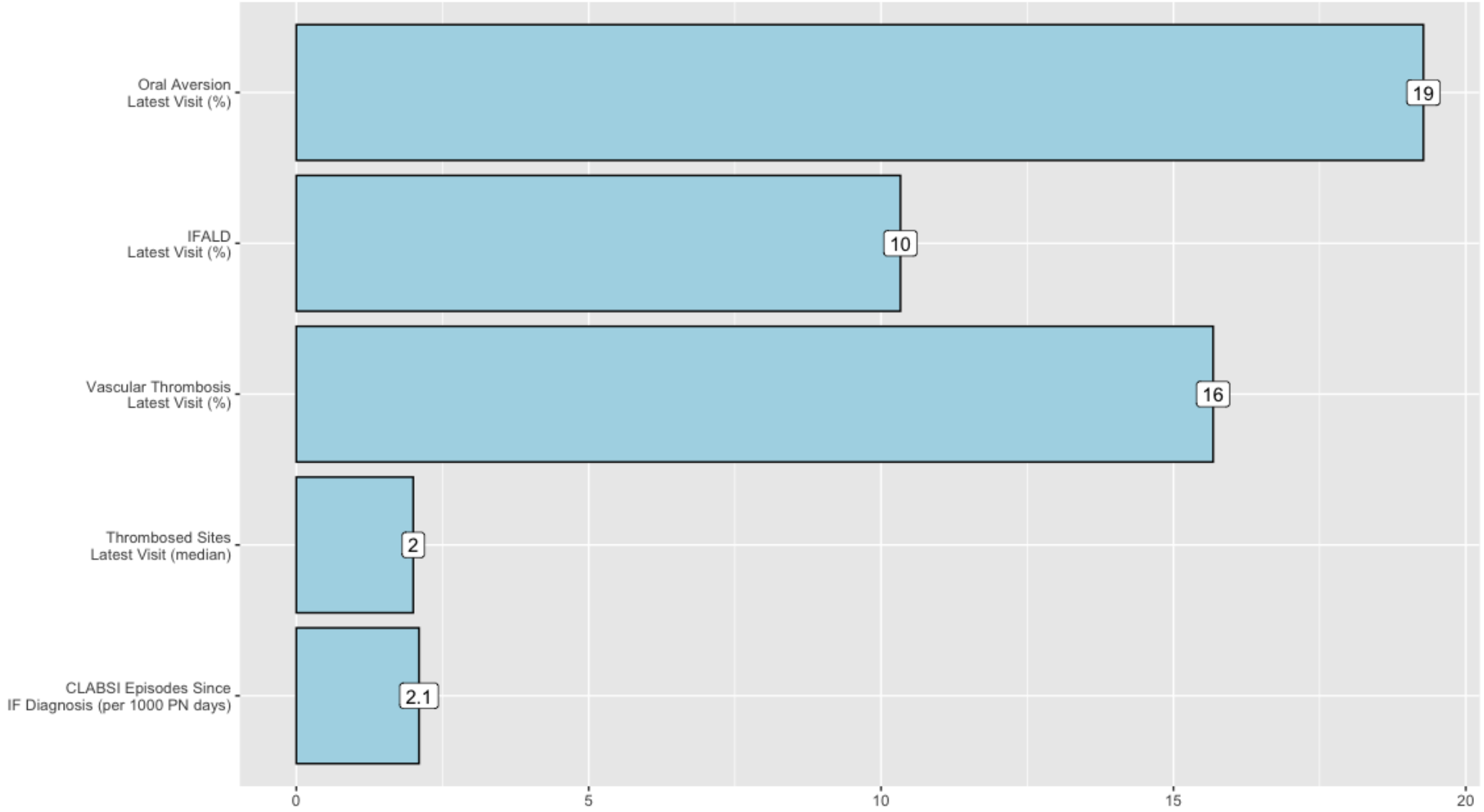
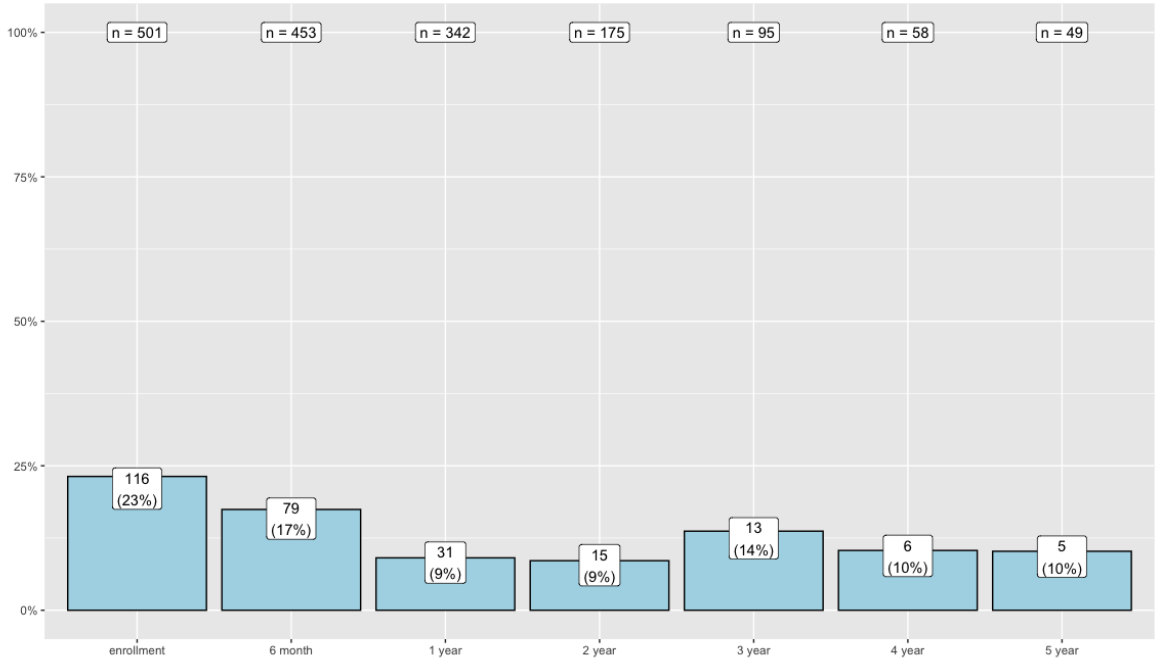
IF Related Complications – Last Visit

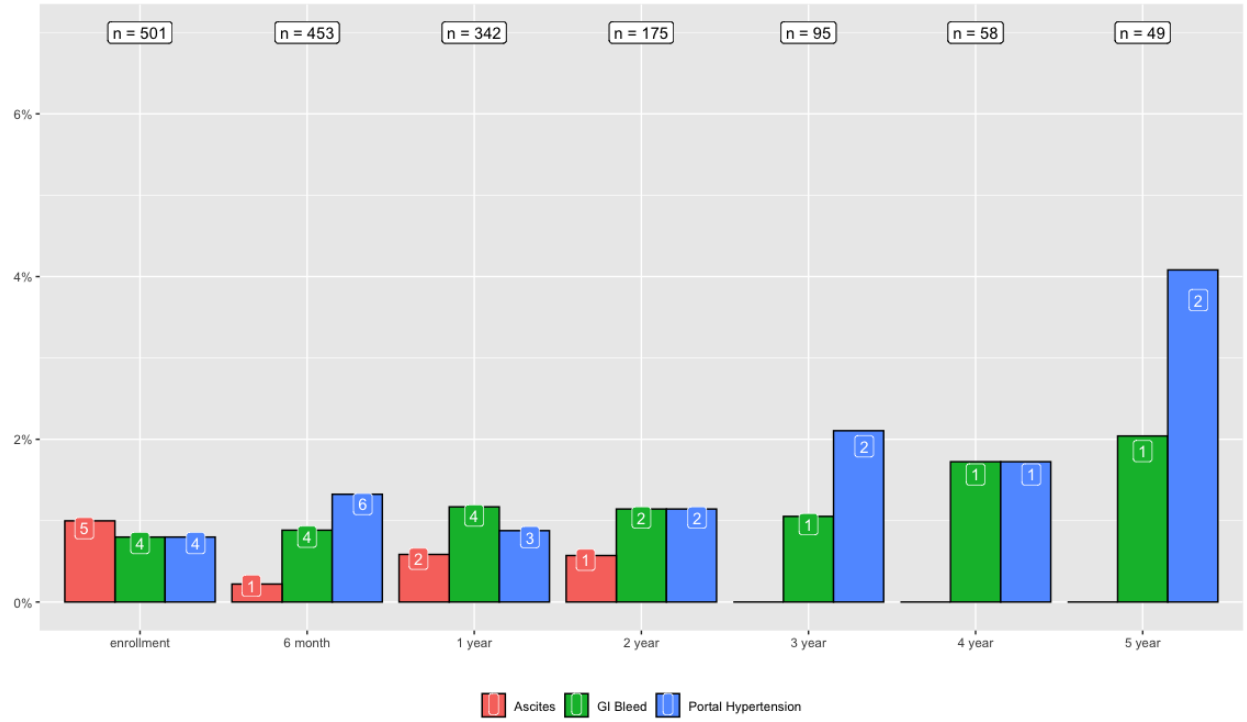


IFALD – Incidence and Complications

IFALD - Incidence



IFALD - Complications



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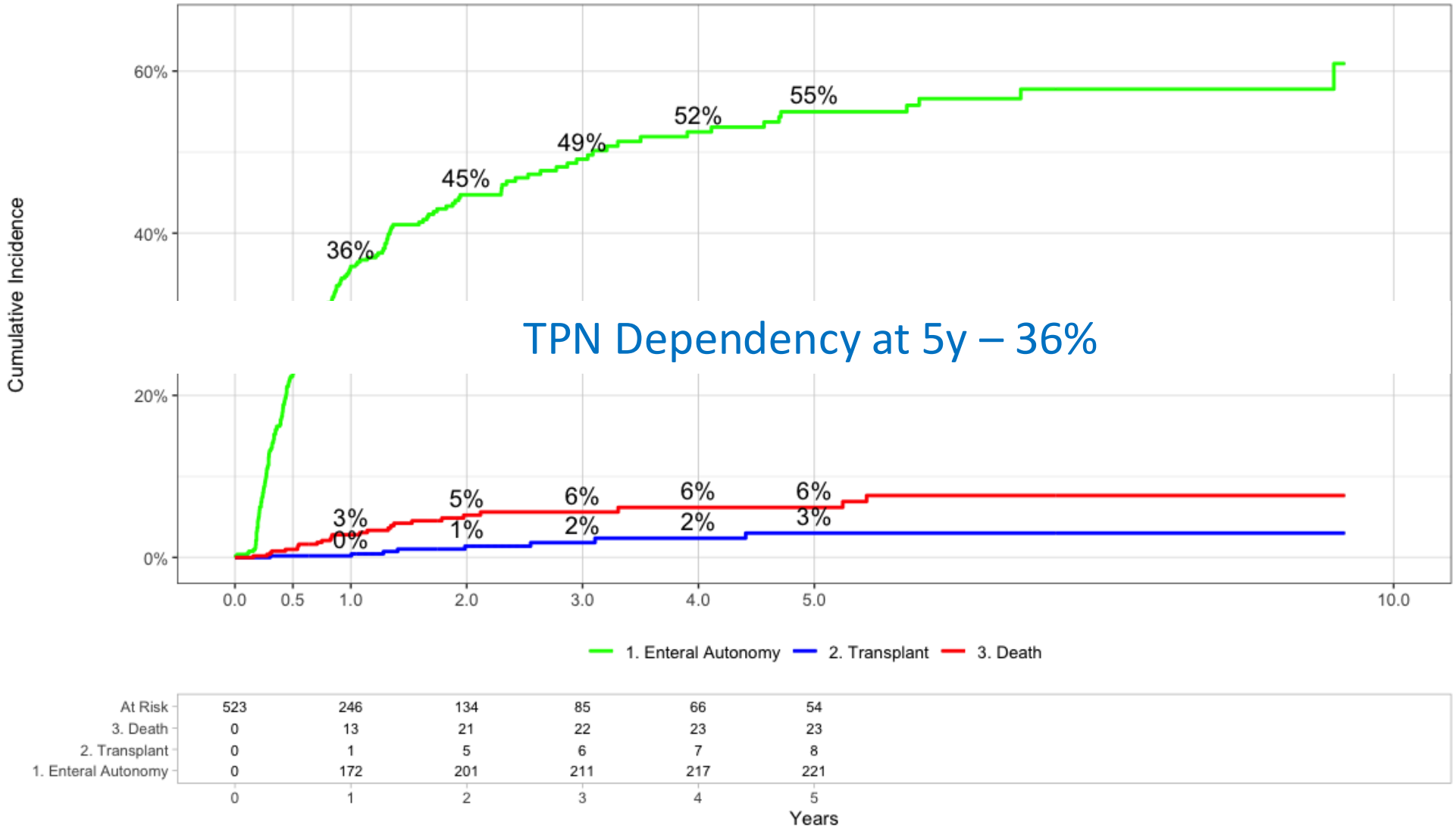
Overall Outcome



TPN Dependency at 5y – 36%

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Future Directions and Priorities

- Identification of beneficial interventions and treatments
- Quality improvement projects through the creation of benchmarking and learning networks
- Utilization of the IIFR large data base for AI and machine learning
- Development of patient reported outcome measures (PROMS) and assessment of quality of life and child development
- Identification of research grants and resources to support the IIFR activities

Summary

- The majority of patients were born prematurely, diagnosed in the first year of life and suffer from SBS with a median of 19% predicted SB length for age
- Enteral nutrition is introduced early in life for most patients and allow TPN weaning over the first year since diagnosis. However, PN calories remain stable for those who do not adapt
- The use of Teduglutide increases with time with 16% of children at 5y post diagnosis
- IFALD is diagnosed in 10% of cases at their last visit and if associated with complications it leads to poor outcome
- Vascular thrombosis and oral aversion are evolving complications that require active management strategies
- CLABSI rate is relatively high at 2.1 infections/1000 PN days. Ideal benchmarking for HPN is required
- Cumulative incidence analysis suggest that 36% remain PN dependent at 5y. A call for research and management efforts to reduce this rate.

Acknowledgments

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Dr. Robert Venick
Dr. Shweta Namjoshi
Dr. Yaron Avitzur

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IIRTA Council

TTS

Joining the IFR

Please contact Dr. Yaron Avitzur and Ms. Nilosa Selvakumaran at nilosa.selvakumaran@sickkids.ca

For more details visit: https://tts.org/2-uncategorised/1089-ifr-irta