

# Evaluation of Inhaled Corticosteroid Use and Indication in Adults with Cystic Fibrosis or CFTR-Related Disorder



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## Background

- Cystic fibrosis (CF) is an autosomal recessive genetic disorder
- Impaired mucociliary clearance increases risk of bacterial infection and airway inflammation, leading to progressive lung function decline
- Inhaled corticosteroids (ICS) are commonly prescribed in people with CF (pwCF) without a definitive indication despite lack of evidence to support this practice

## Objectives

- Primary**
  - To determine the prevalence and indications for ICS use in pwCF and CFTR-related disorder
- Secondary** – to identify:
  - Total daily doses of ICS used
  - Adverse drug reactions (ADRs) attributed to ICS +/- long acting beta agonist (LABA) therapy
  - Drug-drug interactions with ICS therapy
  - Respiratory outcomes related to withdrawal of ICS

## Methods

- Study Design:** Single-centre, retrospective health record review from Nov 9<sup>th</sup>, 2019 to Nov 9<sup>th</sup>, 2020
- Inclusion:** Adults with a diagnosis of CF or CFTR-related disorder at the St. Paul's Hospital CF Clinic
- Exclusion:**
  - Incomplete clinic visit documentation
  - Missing demographics and outcome data
  - < 2 clinic visits per year
  - Lung transplant recipient
- Analysis:** Descriptive statistics

## Figure 1: Reasons for Exclusion

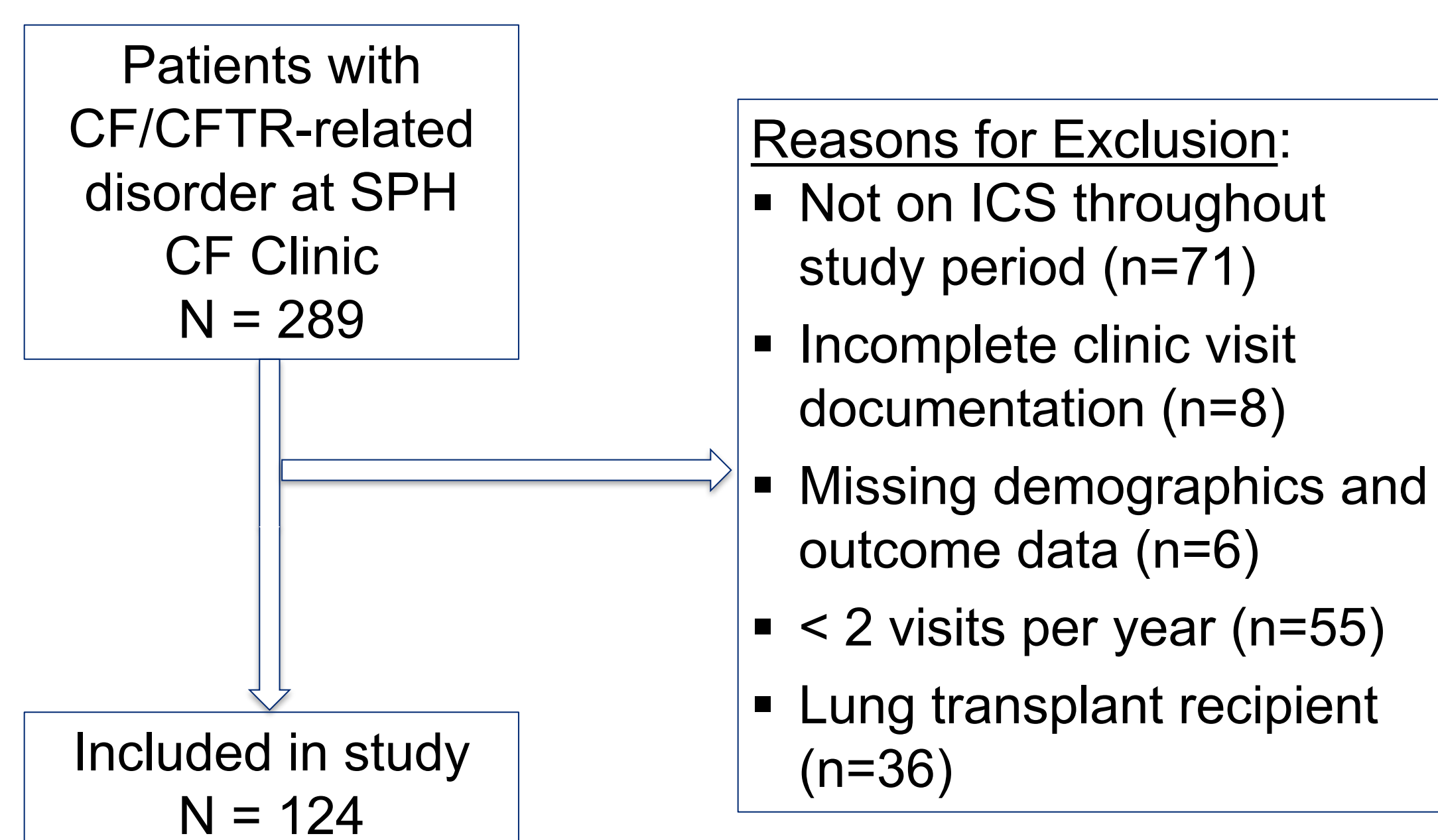


Table 1: Patient Characteristics (N=124)

Age, median [IQR]	36 [27-46]
Sex, % (Male)	48
BMI, median [IQR]	23 [21-25]
Age of CF diagnosis, median [IQR]	2 [0.3-10]
CFTR gene variants, %	
▪ F508d homozygous	56 (45)
▪ F508d heterozygous	46 (37)
▪ Other	6 (4)
▪ CFTR-related disorder	16 (12)
FEV1% predicted, median [IQR]	
▪ Study Entry	61 [46-77]
▪ Study Exit	61 [46-76]
FVC1% predicted, median [IQR]	
▪ Study Entry	84 [71-93]
▪ Study Exit	82 [71-94]

Figure 2: Primary Objective

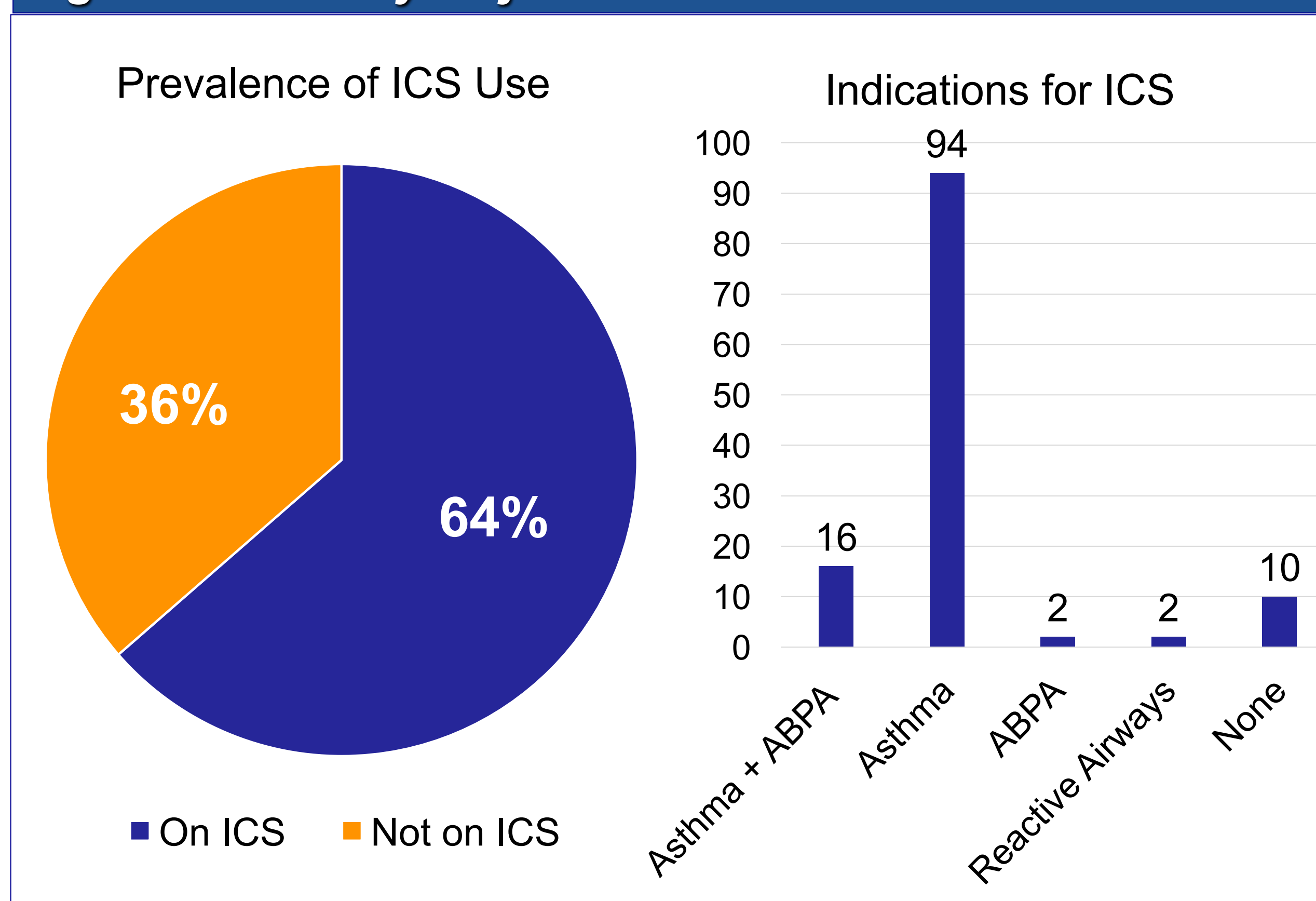
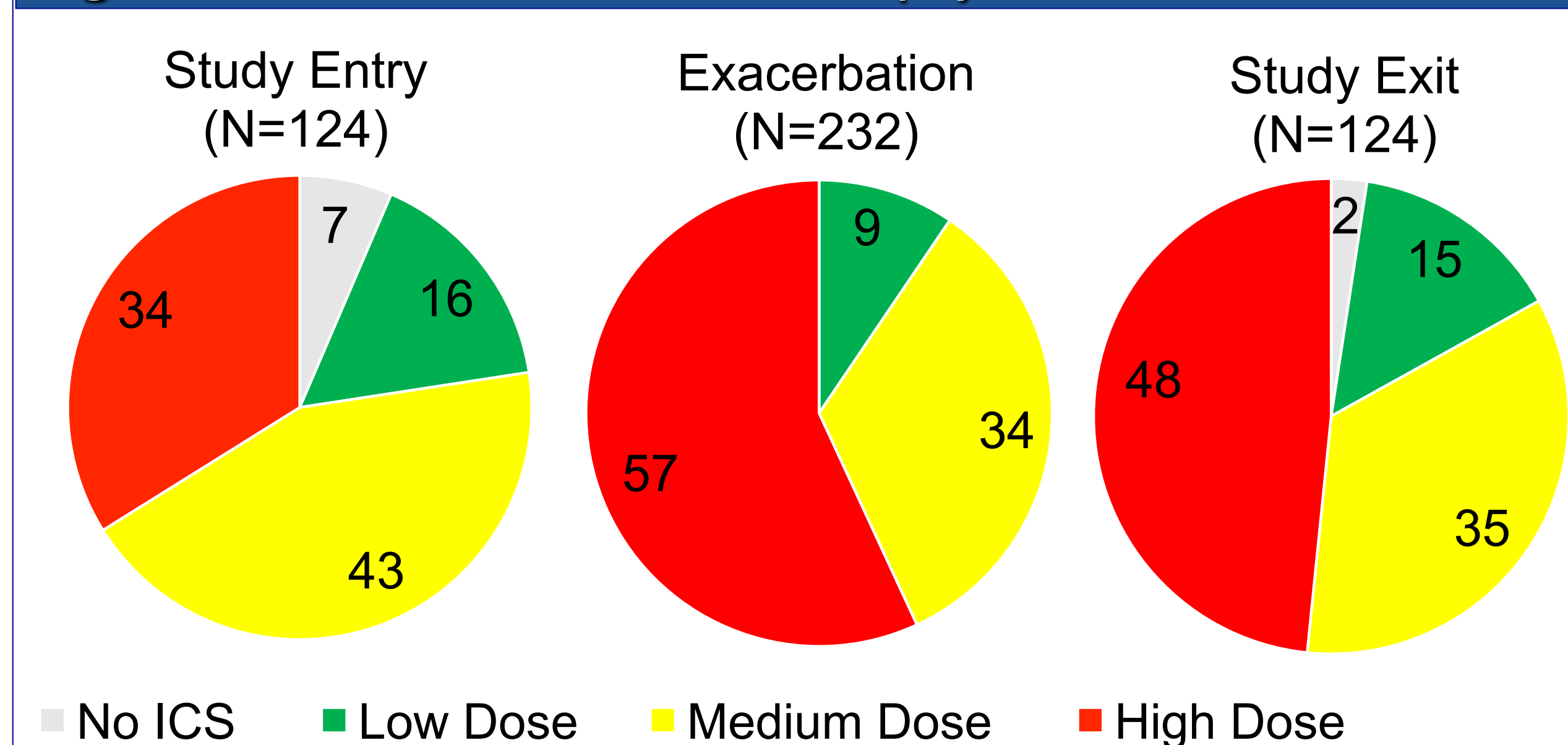


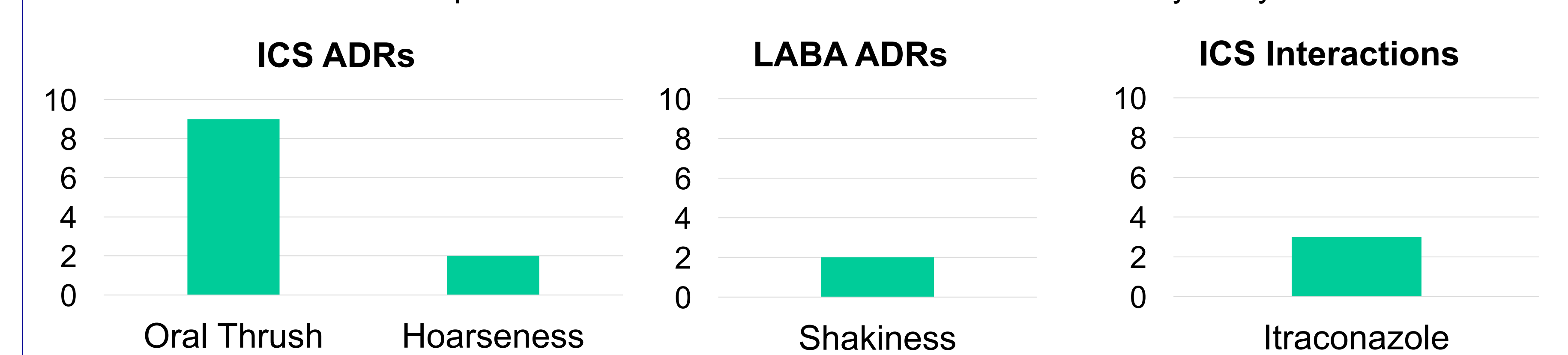
Figure 3: GINA Classifications<sup>1</sup> (%)



## Secondary Objectives

GINA Classifications	Low Dose (mcg/day)		Medium Dose (mcg/day)		High Dose (mcg/day)	
	%	Mean +/- SD	%	Mean +/- SD	%	Mean +/- SD
<b>Study Entry (N<sup>1</sup>=124<sup>3</sup>)</b>						
BUD (n=79)	65	354 +/- 84	90	796 +/- 29	41	1553 +/- 326
MOM (n=13)	10	150 +/- 50	4	350 +/- 50	21	800 +/- 0
FP (n=15)	10	250 +/- 0	6	500 +/- 0	24	1050 +/- 150
FF (n=4)	15	100 +/- 0	0	0 +/- 0	2	200 +/- 0
BEC (n=2)	0	0 +/- 0	0	0 +/- 0	5	800 +/- 0
2 ICS (n=3)	0	N/A	0	N/A	7	N/A
<b>Exacerbation (N<sup>2</sup>=232)</b>						
BUD (n=170)	90	370 +/- 71	91	789 +/- 46	60	1696 +/- 740
MOM (n=32)	5	200 +/- 0	5	375 +/- 43	20	857 +/- 140
FP (n=18)	0	0 +/- 0	4	500 +/- 0	11	1063 +/- 242
FF (n=8)	5	100 +/- 0	0	0 +/- 0	5	214 +/- 35
BEC (n=1)	0	0 +/- 0	0	0 +/- 0	1	800 +/- 0
2 ICS (n=3)	0	N/A	0	N/A	3	N/A
<b>Study Exit (N<sup>1</sup>=124<sup>4</sup>)</b>						
BUD (n=85)	78	357 +/- 82	86	789 +/- 45	56	1641 +/- 455
MOM (n=16)	6	200 +/- 0	9	400 +/- 0	18	818 +/- 134
FP (n=11)	11	175 +/- 75	5	500 +/- 0	12	1071 +/- 175
FF (n=4)	5	100 +/- 0	0	0 +/- 0	5	233 +/- 47
BEC (n=1)	0	0 +/- 0	0	0 +/- 0	2	800 +/- 0
2 ICS (n=4)	0	N/A	0	N/A	7	N/A

1 = # of patients  
 2 = # of episodes  
 3 = number does not add up to 124 due to individuals who did not enter the study on ICS  
 4 = number does not add up to 124 due to individuals who discontinued ICS by study exit



## Limitations

- Reliant on accuracy of documented data
- When ranges of ICS were used, the higher dose was recorded
- CF clinic transitioned to virtual clinics during COVID-19 pandemic; therefore, PFTs not consistently available
- Potential misclassification of ICS/LABA-related ADRs, if secondary to unrelated cause

## Conclusions

- At our adult centre, 64% of pwCF or CFTR-related disorder were on ICS, of whom 92% had a diagnosis of asthma, Allergic Bronchopulmonary Aspergillosis (ABPA), and/or reactive airways
- Insufficient data to evaluate impact of ICS withdrawal on lung function and safety outcomes
- Observed increase in average ICS dose during times of pulmonary exacerbation and by time of study exit

## References:

1. Guide GP. Pocket guide for asthma management and prevention for adults and Children older than 5 years. A pocket guide for Health Professionals. 2019.