



CF Transition

Graduating from Pediatric to Adult Care



The Face of CF





CF requires multidisciplinary care

Manifestations of Cystic Fibrosis

- General**
- Growth failure (malabsorption)
 - Vitamin deficiency states (vitamins A, D, E, K)

- Nose and sinuses**
- Nasal polyps
 - Sinusitis

- Liver**
- Hepatic steatosis
 - Portal hypertension

- Gallbladder**
- Biliary cirrhosis
 - Neonatal obstructive jaundice
 - Cholelithiasis

- Bone**
- Hypertrophic osteoarthropathy
 - Clubbing
 - Arthritis
 - Osteoporosis

- Intestines**
- Meconium ileus
 - Meconium peritonitis
 - Rectal prolapse
 - Intussusception
 - Volvulus
 - Fibrosing colonopathy (strictures)
 - Appendicitis
 - Intestinal atresia
 - Distal intestinal obstruction syndrome
 - Inguinal hernia

- Lungs**
- Bronchiectasis
 - Bronchitis
 - Bronchiolitis
 - Pneumonia
 - Atelectasis
 - Hemoptysis
 - Pneumothorax
 - Reactive airway disease
 - Cor pulmonale
 - Respiratory failure
 - Mucoid impaction of the bronchi
 - Allergic bronchopulmonary aspergillosis

- Heart**
- Right ventricular hypertrophy
 - Pulmonary artery dilation

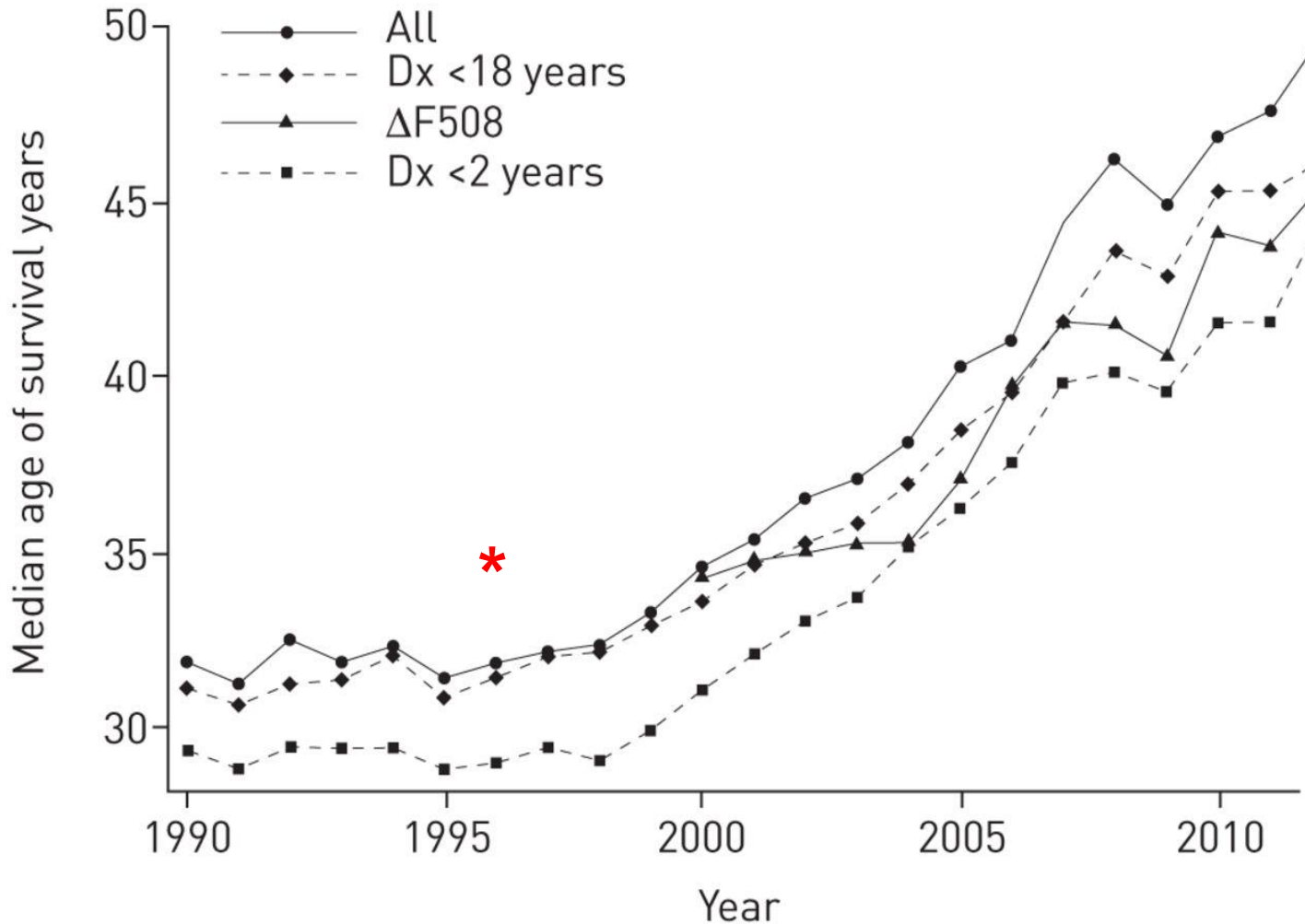
- Spleen**
- Hypersplenism

- Stomach**
- GERD

- Pancreas**
- Pancreatitis
 - Insulin deficiency
 - Symptomatic hyperglycemia
 - Diabetes

- Reproductive**
- Infertility (aspermia, Absence of vas deferens)
 - Amenorrhea
 - Delayed puberty

The Scope of the Problem



In 1996, The CF Foundation recommended that all CF Centers transition at least 90% of their patients aged ≥ 21 years to adult providers.

The New Faces of CF



Transition: A period or process of changing from one state or condition to another

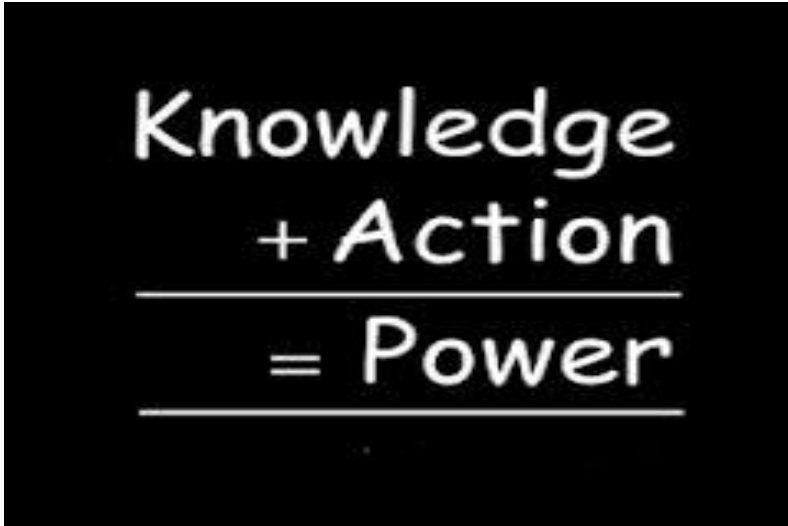
- Barriers
 - » Logistics
 - » Parents not “letting go”
 - » Concerns over quality of care and poor communication
 - » Family-focused care limits education, independence, and self-management skills for the adolescent
- Consequence
 - » Patients not seeking care until a serious problem arises

Chaudhry SR et al. *Ped pulm.* 2013;48(7):658-65.
Nasr SZ et al. *J adol health.* 1992;13(8):682-5.
Blum RW. *J adol health.* 1995;17(1):3-5.

Flume PA. *Curr opin pulm med.* 2009; 15(6):611-4.
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Rosen D. *J adol health.* 1995;17(1):10-6.

Project Aim

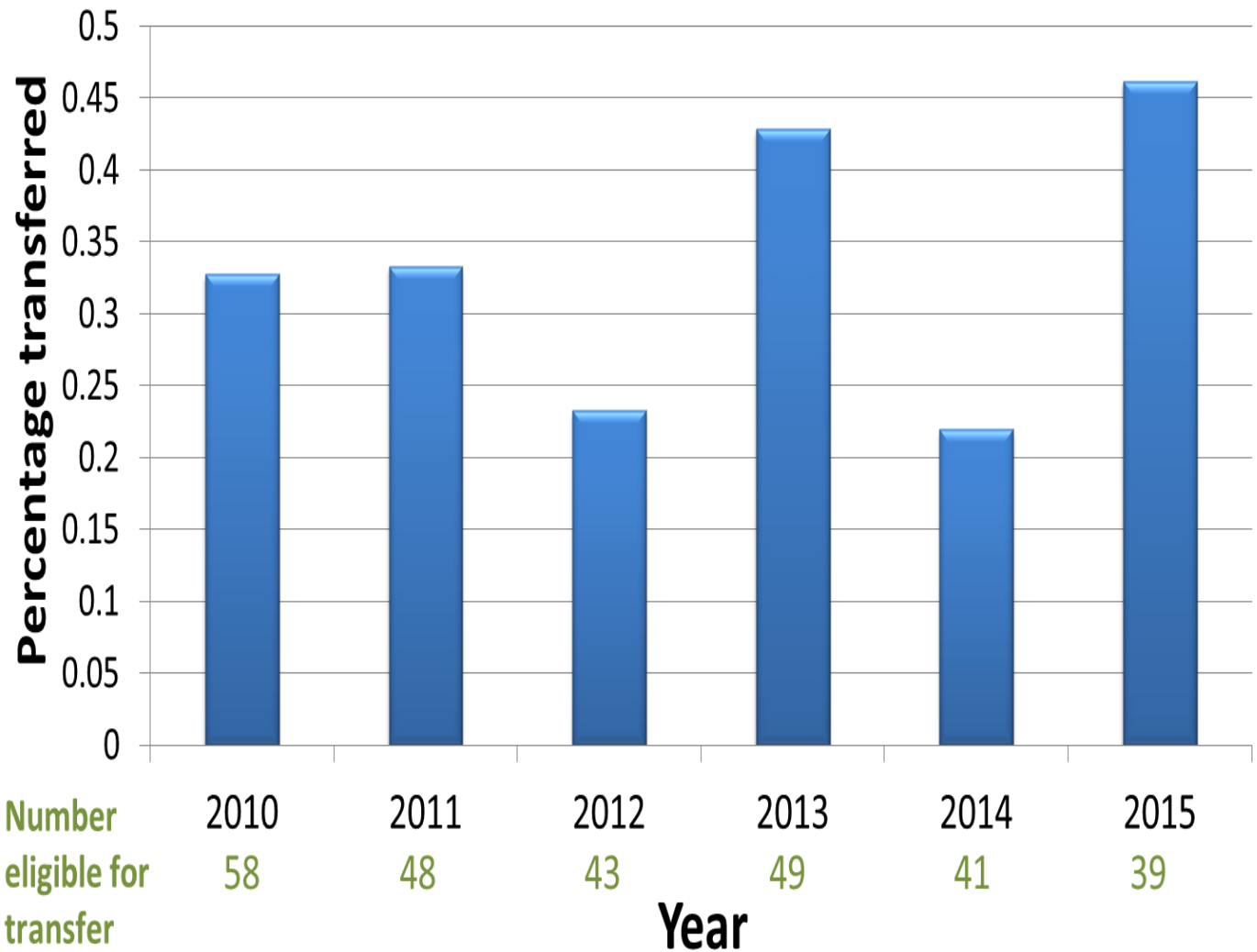
To improve the CF transition process at UNC by creating a sustainable, streamlined, systematic approach to transition for patients beginning early in adolescence and extending to the hand off to adult clinic (age ≥ 18). We expect this will improve health literacy among patients, reduce the number of delayed CF transitions, and improve patient and provider satisfaction.



Knowledge
+ Action
= Power

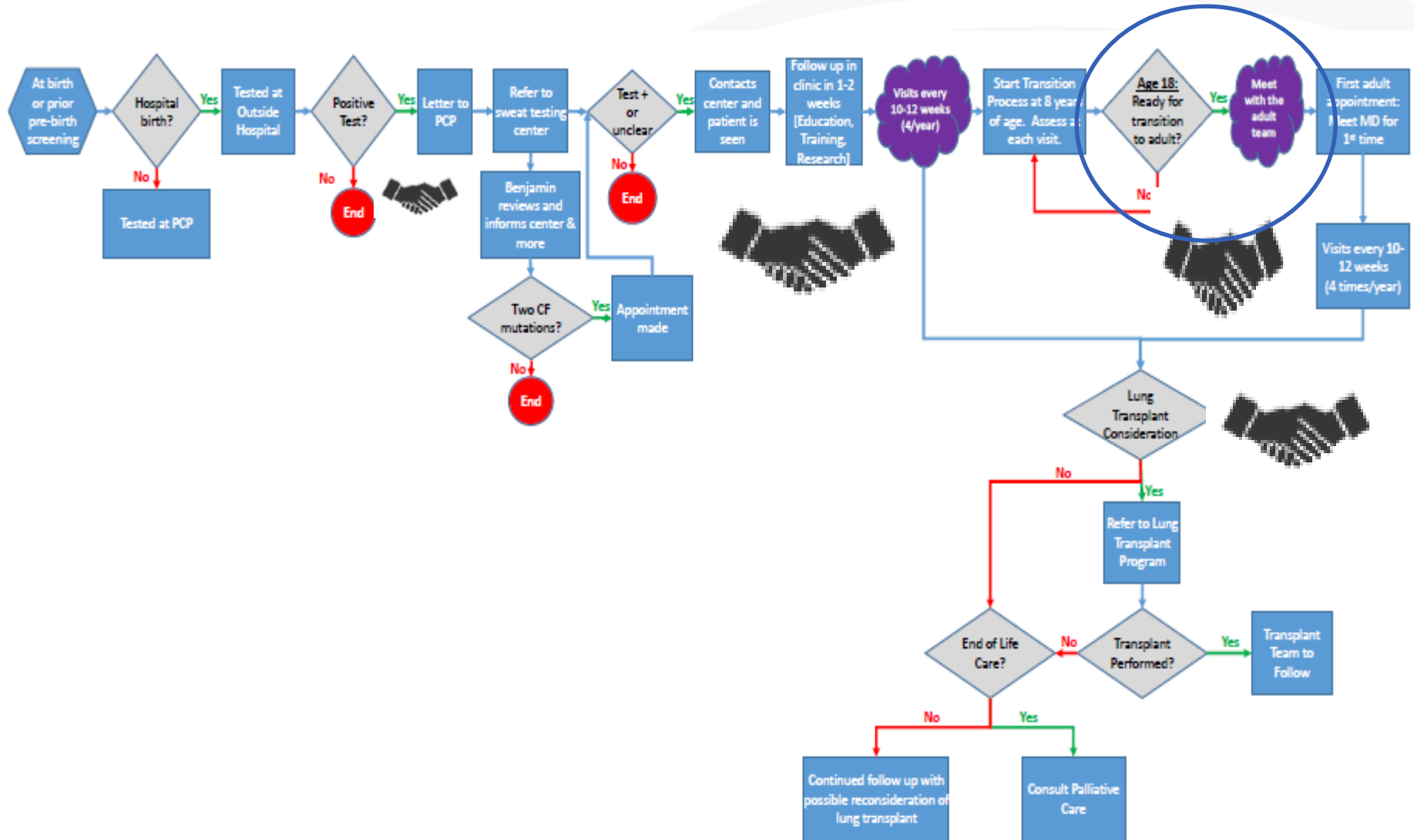
Know your 5 P's

Percent of Eligible Patients Transferred





Process Flow Map



New Process Led by Transition Coordinator



CF R.I.S.E.

LUNG HEALTH & AIRWAY CLEARANCE

THIS INFORMATION MEETS THE GUIDELINES AND STANDARDS OF THE CYSTIC FIBROSIS FOUNDATION'S EDUCATION COMMITTEE.

CF R.I.S.E.
Responsibility. Independence. Safe Care. Education.

LUNG HEALTH & AIRWAY CLEARANCE

Name: _____ Note: This is not a test. This assessment has been developed to help you become more knowledgeable about your CF, with the help of your CF care team.
Date: _____

This assessment is to be completed by a person who is 16 or older with cystic fibrosis (CF). For each question, please read all the answer choices carefully before choosing the **one** answer you think is **best**. If you don't know an answer, leave it blank and move on to the next question.

*1. Pulmonary function tests and spirometry are a fancy way of saying:
a) Show how your lungs are working
b) Only need to be done once in a while
c) Depend on your effort in blowing
d) Answers a and c

*2. Which of the following is the maximum amount exhaled from the lungs the deepest breath possible?
a) Forced expiratory volume
b) Forced vital capacity (FVC)
c) Both FEV1 and FVC are spirometry
d) All of the above

*3. Timing is everything. To take the following to:
a) Airway clearance, inhaled short-acting bronchodilators
b) Inhaled short-acting bronchodilators, and inhaled antibiotics, inhaled bronchodilators, and airway clearance
c) Inhaled antibiotics, inhaled bronchodilators, and airway clearance
d) Airway clearance, inhaled short-acting bronchodilators, and inhaled antibiotics

*4. Environmental factors cause additional lung damage.
a) True
b) False

*5. All of the CF bacteria in your lungs will be killed with IV antibiotics.
a) True
b) False

*6. How can you decrease the number of lung infections?
a) Stronger heart muscle
b) Potential improvements in lung function
c) Being in a better mood
d) All of the above

7. Regular exercise can result in:
a) stronger heart muscle
b) Potential improvements in lung function
c) Being in a better mood
d) All of the above

8. The following are symptoms of a pulmonary "exacerbation":
a) Increased cough
b) Decreased lung function
c) Increased appetite
d) Answers a and b
e) All of the above

CF R.I.S.E. was developed in collaboration with a multidisciplinary team of CF experts and is sponsored by Gilead.

11 Assessment Topics

- CF Liver Disease (CFLD)
- CF-Related Diabetes (CFRD)
- College & Work
- Equipment Maintenance & Infection Control
- Sexual Health
- General CF Health
- Insurance & Financial
- Lifestyle
- Lung Health & Airway Clearance
- Pancreatic Insufficiency & Nutrition
- Screening & Prevention

CF Responsibilities Checklist

3: Living With Cystic Fibrosis


Name: _____

Note: There are no right or wrong answers to this survey.

Date: _____

Please provide your honest feedback below so that we can work together to improve the management of your CF over time.

1	2	3	4	5	NA
I am completely responsible	I am primarily responsible	My support person and I are equally responsible	My support person is primarily responsible	My support person is completely responsible	Not Applicable

Please indicate the person in your household responsible for each of the following healthcare behaviors by placing the appropriate number from 1 to 5, or NA for Not Applicable, in the boxes below: 

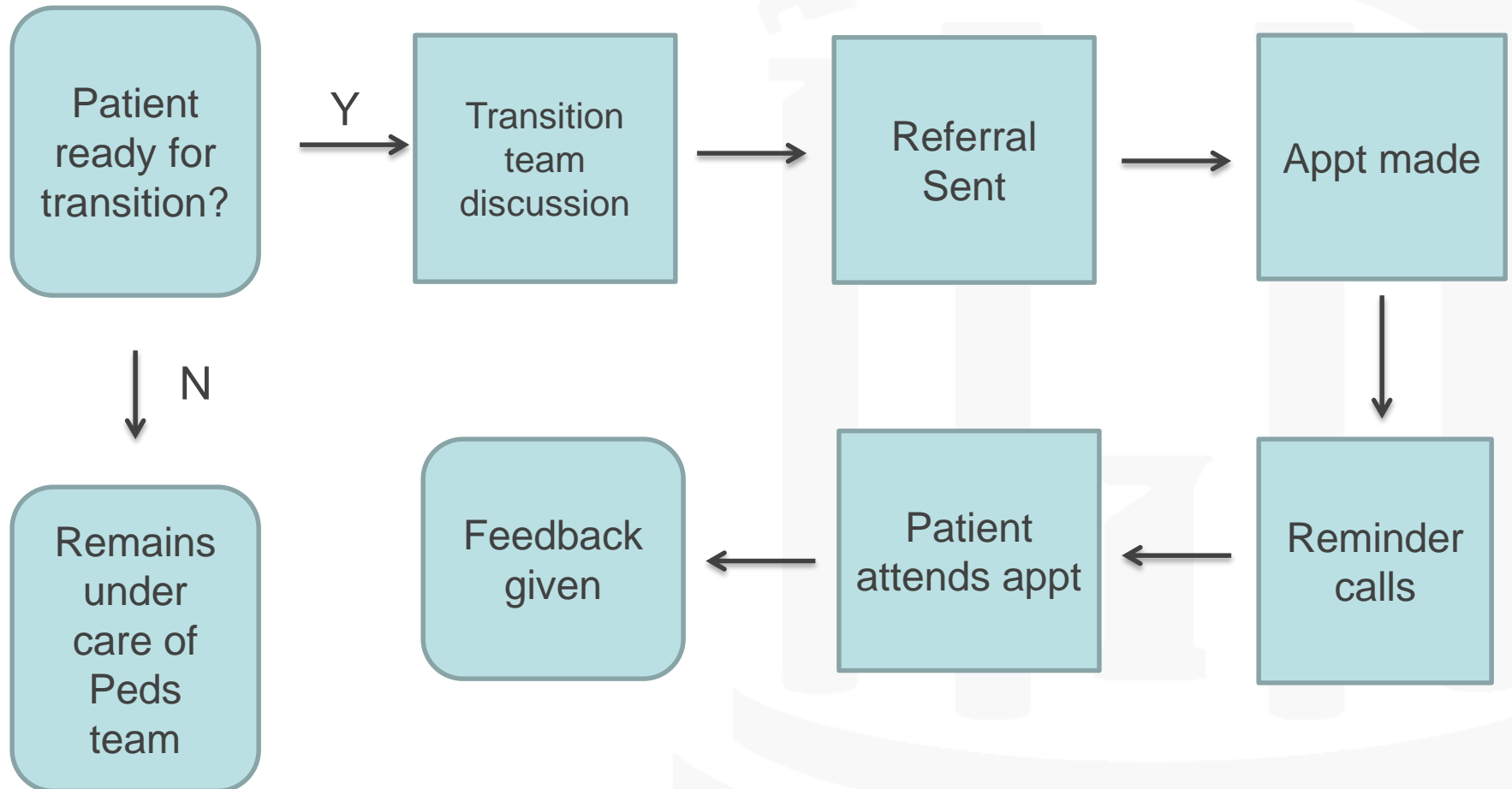
1. Identifying someone with whom psychological and emotional issues can be discussed	
2. Telling close friends, family members, teachers, administrators, etc. about CF	
3. Knowing how to answer or having prepared answers for questions about CF from others	
4. Making sure to follow infection control standards (eg, cleaning your nebulizer regularly)	
5. Making healthy lifestyle choices related to drugs, alcohol, and cigarettes	
6. Recognizing limits (eg, days you may need extra sleep)	
7. Making time for exercise	
8. Making time for fun and hobbies	
9. Preparing for hospital visits/tune-ups	

Add all individual numbers entered for each row above and insert the total on the line below. Divide the total by 9 and enter the result in the box.



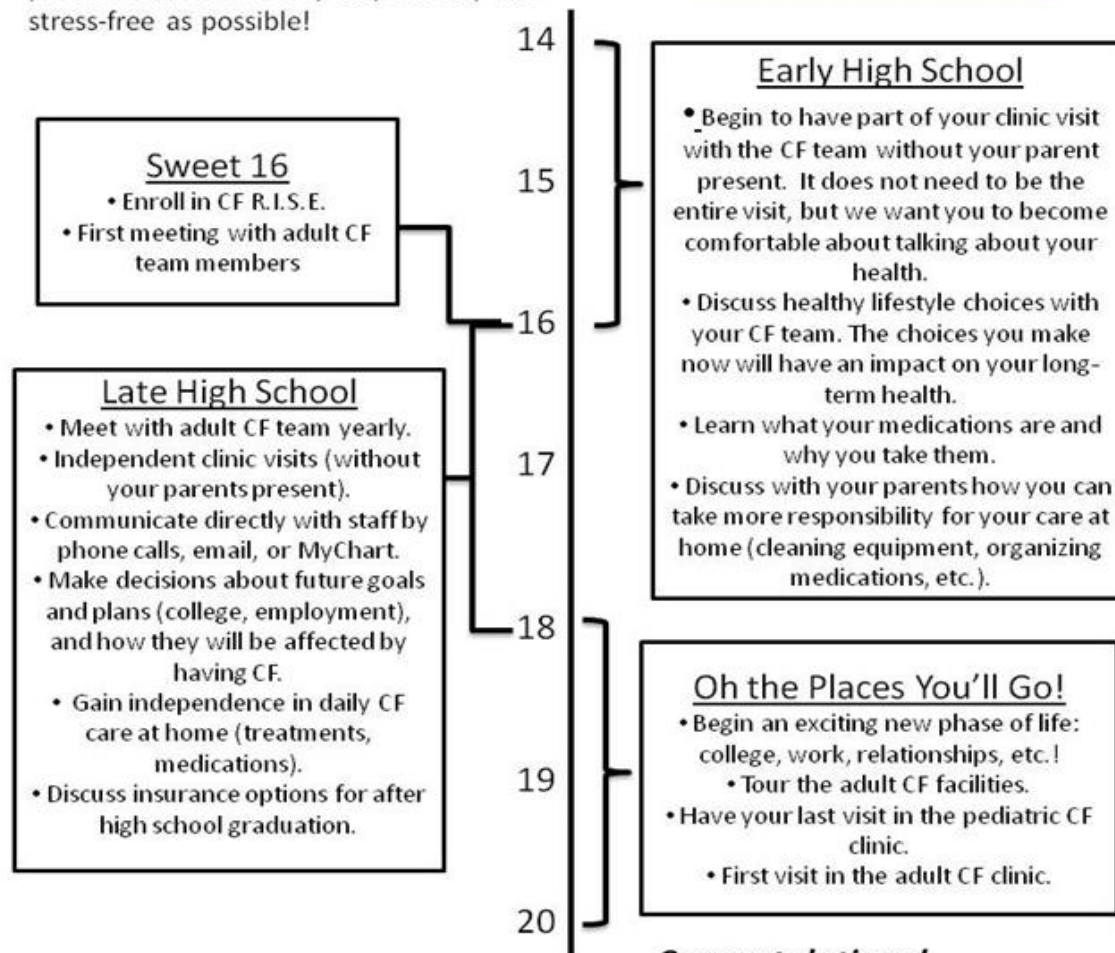


Flow Chart for Patient Transition



CF Transition Pathway

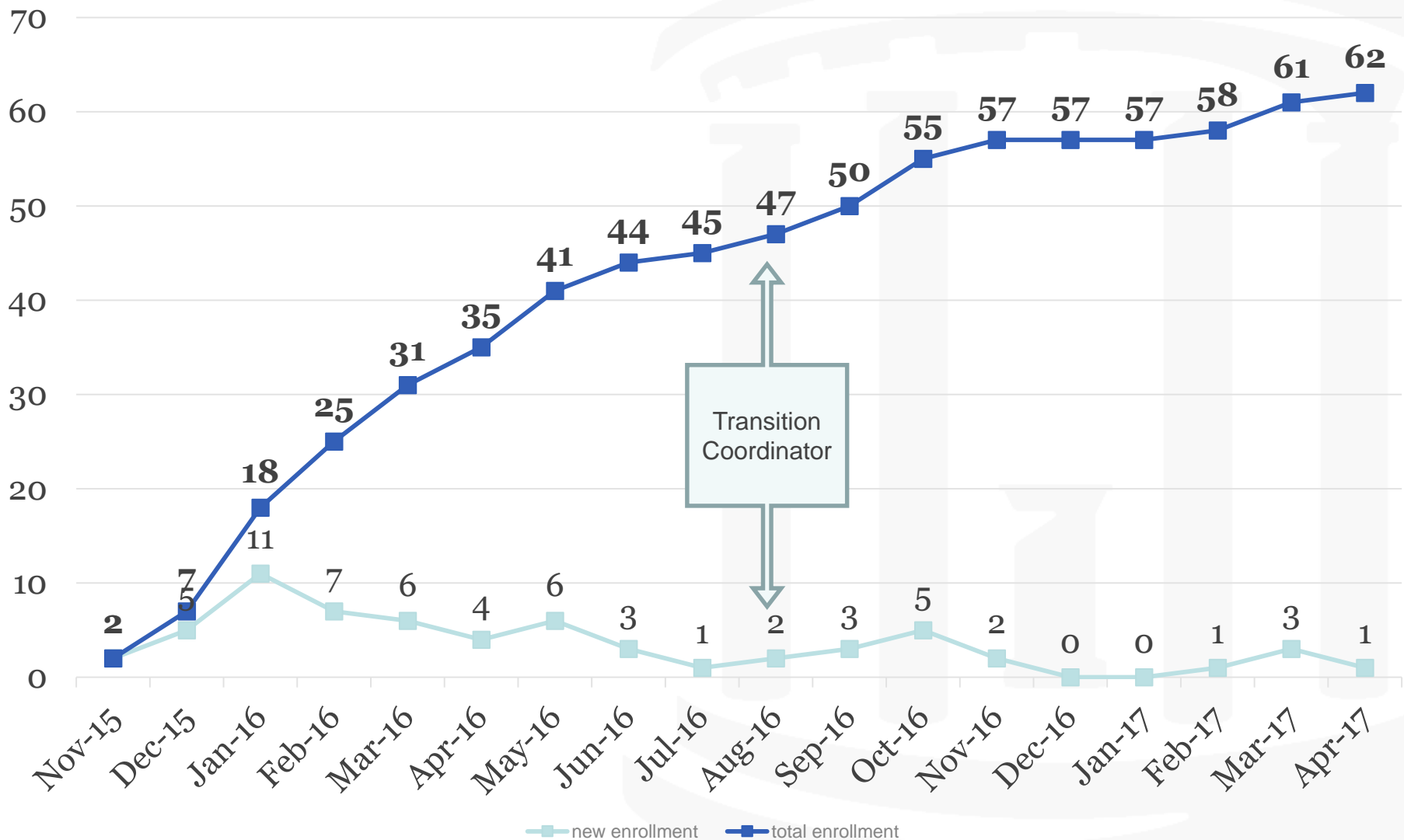
You're entering a very exciting phase of life! You have been learning a lot about what it means to have CF as you've gotten older, and we will continue to work with you to learn everything you need to know to be an adult living with CF. Part of being an adult with CF is transitioning to the adult CF care team. Don't panic! We're here to usher you along in this process and make every step as easy and stress-free as possible!



Congratulations!

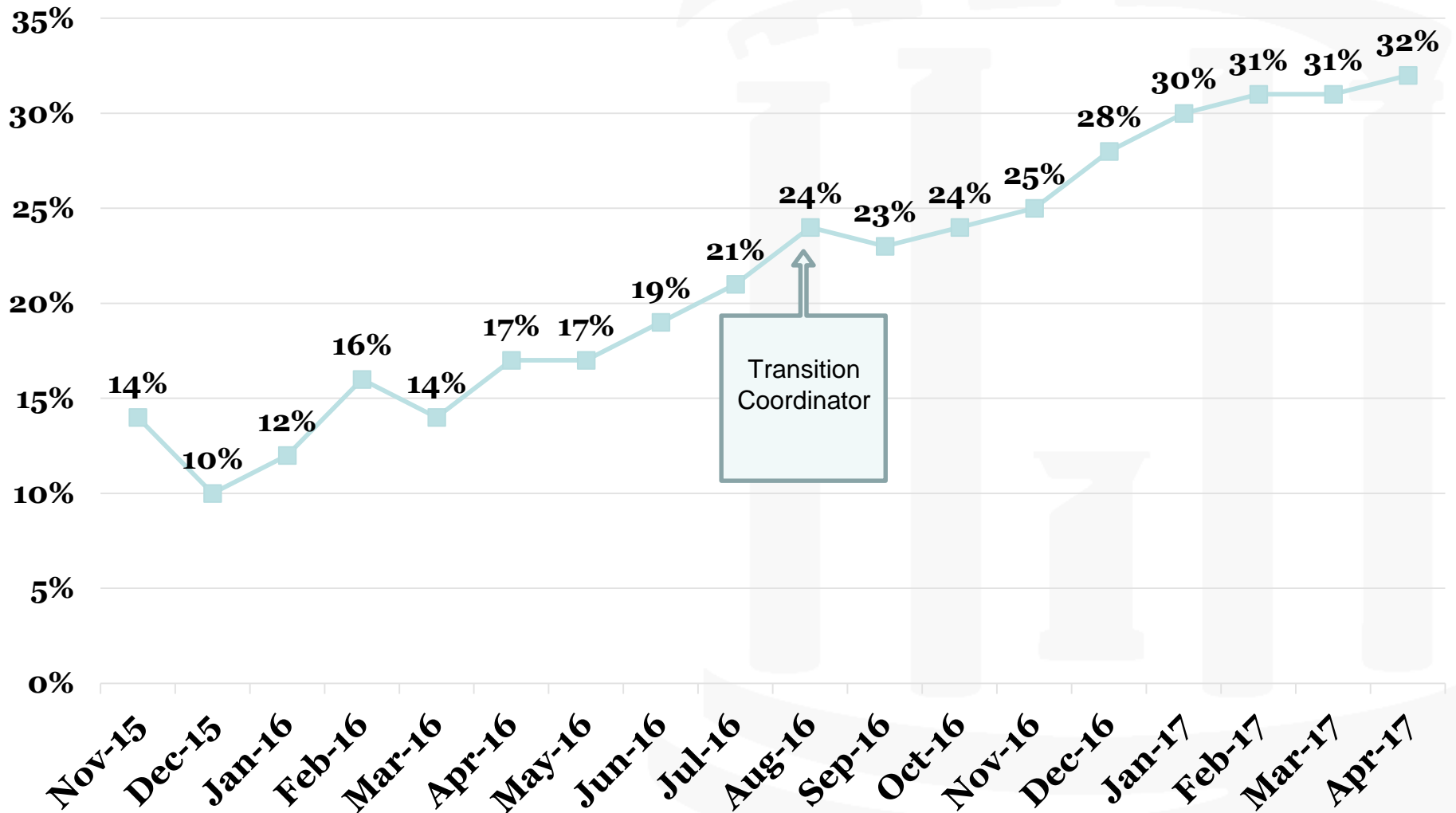


CF R.I.S.E. Enrollment



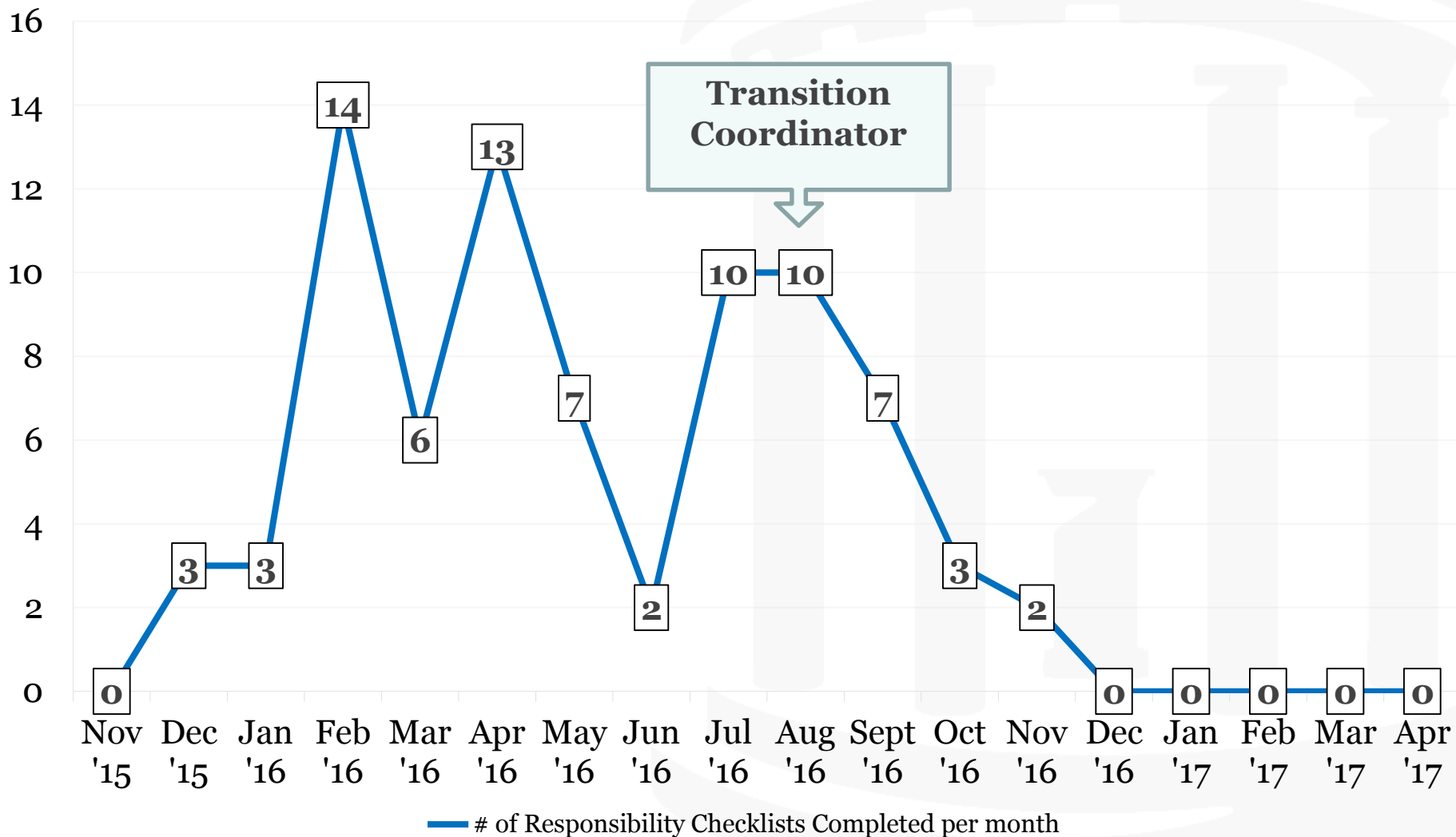


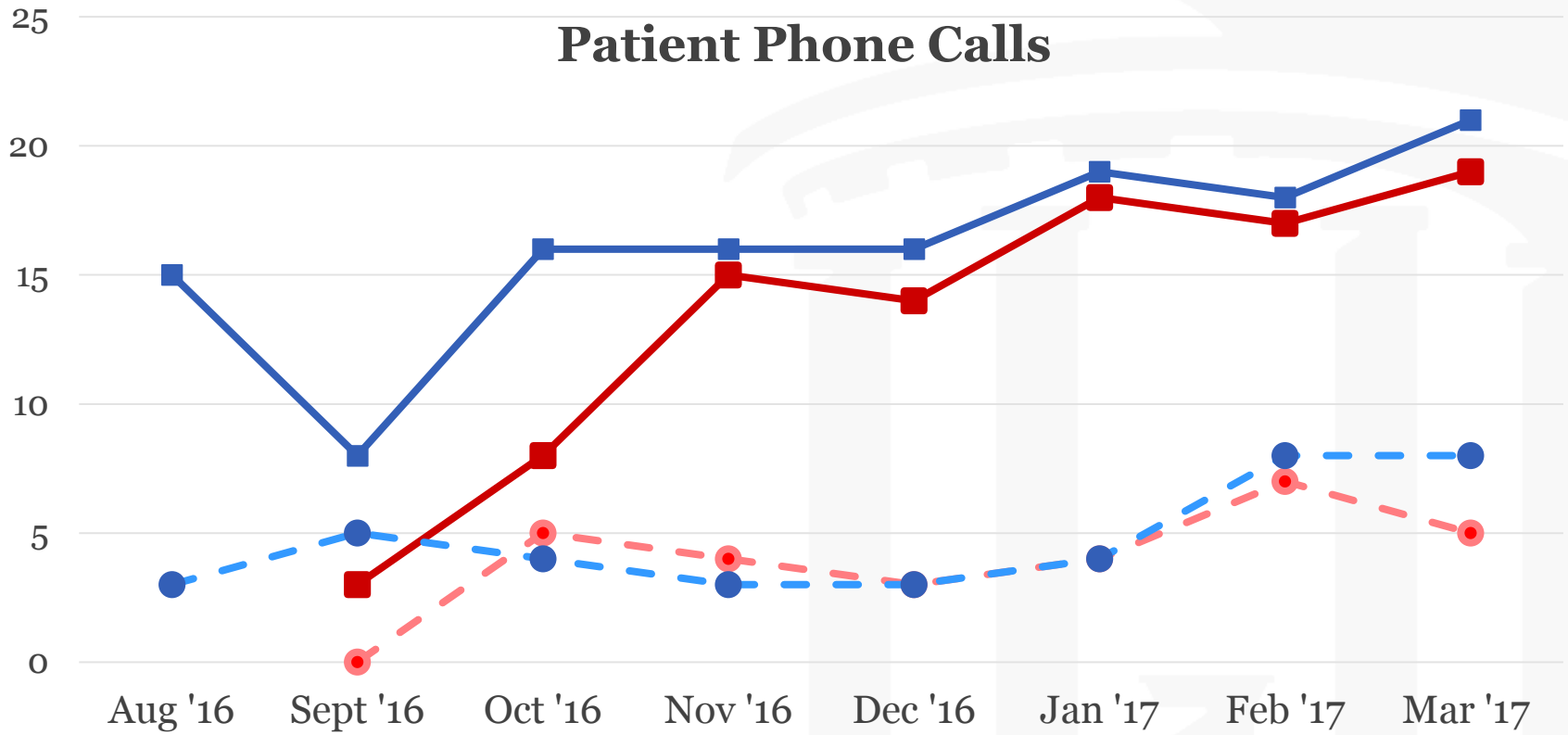
Percent of Knowledge Assessments Completed





CF R.I.S.E. RESPONSIBILITY CHECKLISTS COMPLETED





- Number of pre-visit phone calls made
- number of successful pre-visit phone call attempts
- Number of follow-up calls made
- Number of successful follow-up call attempts

A Patient's Story

- Looking to live independently from parents
- Completed the CF R.I.S.E. insurance & finances knowledge module and set transition goals
- **Patient:** “I feel much better about living by myself now. It is the first time I really thought about the financial side of my disease”.
- **Mother:** “My mind is more at ease now. I feel better about letting my child go.”

Sustainability Plan

Transition
Coordinator

Streamline
Approach

Implementation
Guide/Toolkit

Track
Outcomes



Spread Plan

- **Institutionally**
 - » Other chronic childhood diseases
 - Pulmonary focus at first (asthma, PCD, etc.)
 - Primary Care Provider
 - » Implementation toolkit and role descriptions
- **Nationally**
 - » CF R.I.S.E. for age range >10
 - » ATS working group
 - » Got Transition and ACP Resources for Transition



Lessons Learned

- Program Champion(s)
- Institutional Buy-in
- Patient/family engagement
- Minimize work for others



Acknowledgements

- CF Transition Team
- Pediatric and Adult CF Teams
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 - » Jacob Reardon



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