

Identifying Barriers to Oral Iron Chelator Adherence in Children and Adolescents with Sickle Cell Disease

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Introduction

Exjade is an iron oral chelator necessary in patients with sickle cell disease (SCD) who have received chronic red blood cell transfusions and develop transfusional iron overload, placing the body at risk for iron damage and end-organ failure. A significant body of evidence exists that delineates barriers to iron chelation in patients with thalassemia, however, less data is available regarding barriers to iron chelator adherence in patients with SCD.

Objectives

- Identify barriers to Exjade adherence and determine most common reasons for missed doses of Exjade in patients with SCD treated at Children’s Healthcare of Atlanta at Hughes Spalding’s AFLAC Hematology Clinic (CHOA-HS).
- Identify trends in lab values versus patient reported adherence across the study population.
- Query patients and caregivers for recommendations for improving Exjade adherence.

Methods

- Cross-sectional analysis** with a goal to recruit a convenience sample of 20 patients from CHOA-HS; actual cohort included 18 patients.
- Inclusion Criteria:** SCD, 5 – 18 years old, receive chronic RBC transfusions currently or in the past and are currently prescribed Exjade.
- Exclusion Criteria:** patients with idiopathic hemochromatosis, chronic erythrocytapheresis, and inability to understand English.
- Survey Tool:** Patients were given the Brief Medication Questionnaire (BMQ) and Caregiver Questionnaires (CQ) were also used to evaluate barriers and solicit recommendations.
- Chart Review:** Data collected from EMR for comparison of reported adherence with lab values.

Results

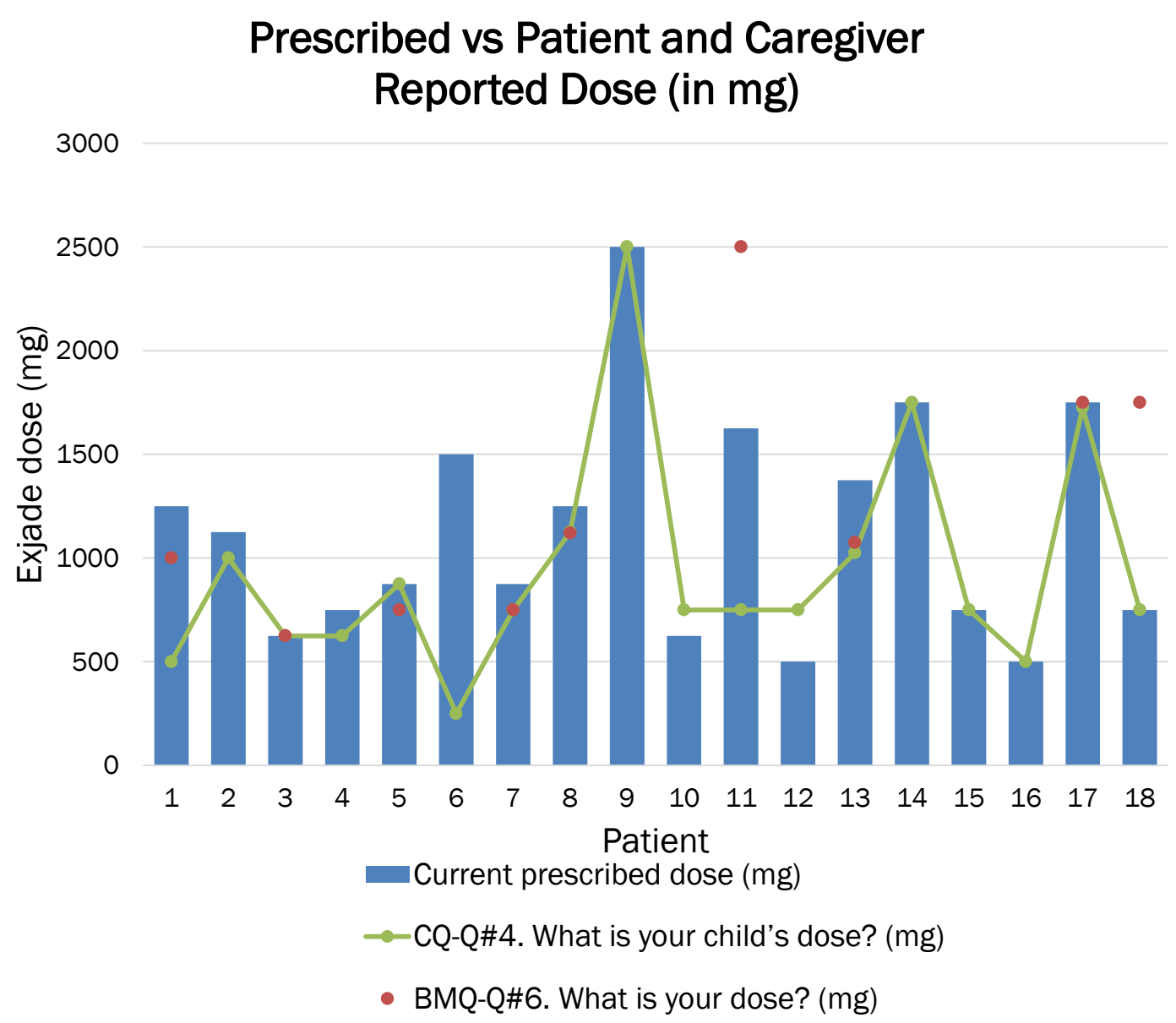


Chart 1. Data from the BMQ-Q#6 and CQ-Q#4 compared to prescribed doses as documented in the EMR revealed 47% of caregivers gave an accurate dose and 18% of patients gave an accurate dose in mg.

Table 1. Demographics		n=18	N%
Subject Age Group			
Child (5-9yo)		6	33%
Preteen (10-12yo)		7	39%
Adolescent (13-18yo)		5	28%
Subject Gender			
Male		8	44%
Female		10	56%
Duration of Therapy			
Years on Chronic Transfusion (mean and SD for 12 of 18 participants)		5	±2.4
Years on Exjade (mean and SD for 11 of 18 participants)		3	±0.4
CQ-Q#6: Reported highest level of education caregiver completed			
College or higher		11	61%
Grade level (K-12)		6	33%
No Response		1	6%
CQ-Q#9b: Does your household income meet family's needs?			
Yes		15	83%
No		2	11%
No Response		1	6%
CQ-Q#9c: Does your child with sickle cell disease have insurance coverage?			
Yes		16	89%
No		1	6%
No Response		1	6%
Type of Insurance			
Medicaid		12	67%
Peachstate		3	17%
Amerigroup		2	11%
Wellcare		1	6%

Table 1. The 18 study participants were stratified by age into cohorts of child, preteen and adolescent; and by gender. Mean years on transfusion therapy was 5±2.4 and years on Exjade 3±0.4. All were covered by some form of Medicaid per EMR.

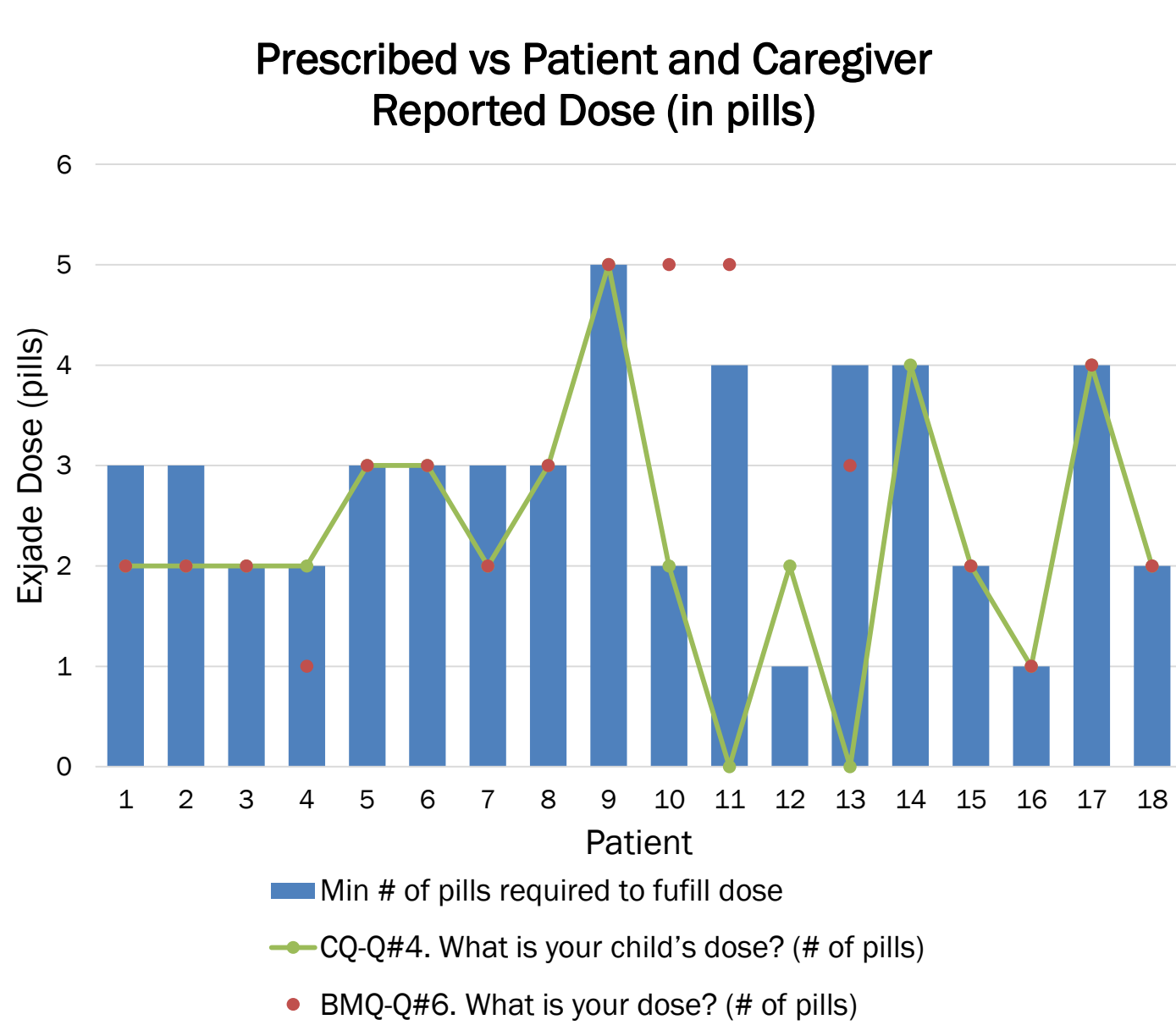


Chart 2. Data from the BMQ-Q#6 and CQ-Q#4 compared to prescribed doses as documented in the EMR revealed 75% of caregivers gave an accurate dose and 56% of patients gave an accurate dose in number of pills.

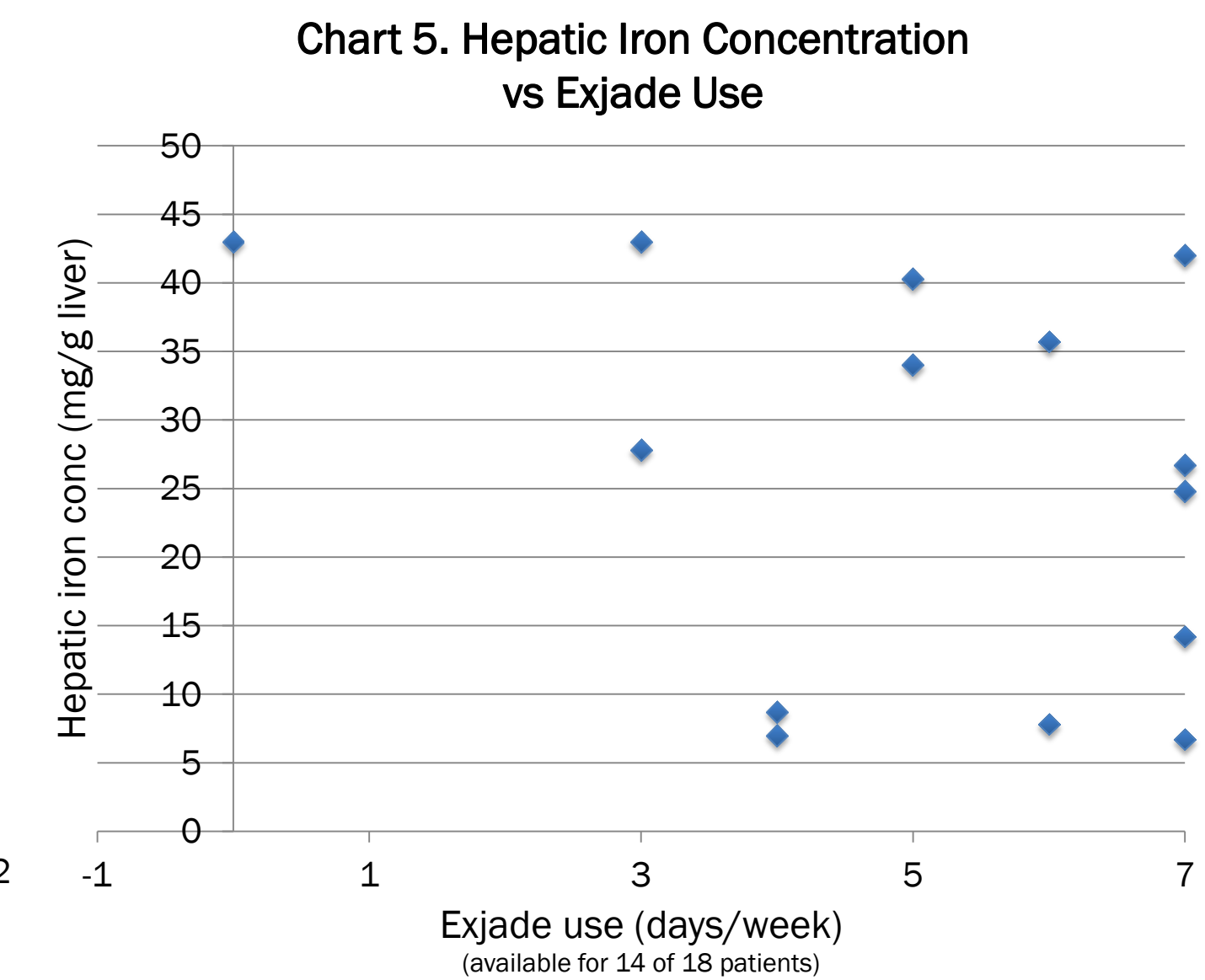
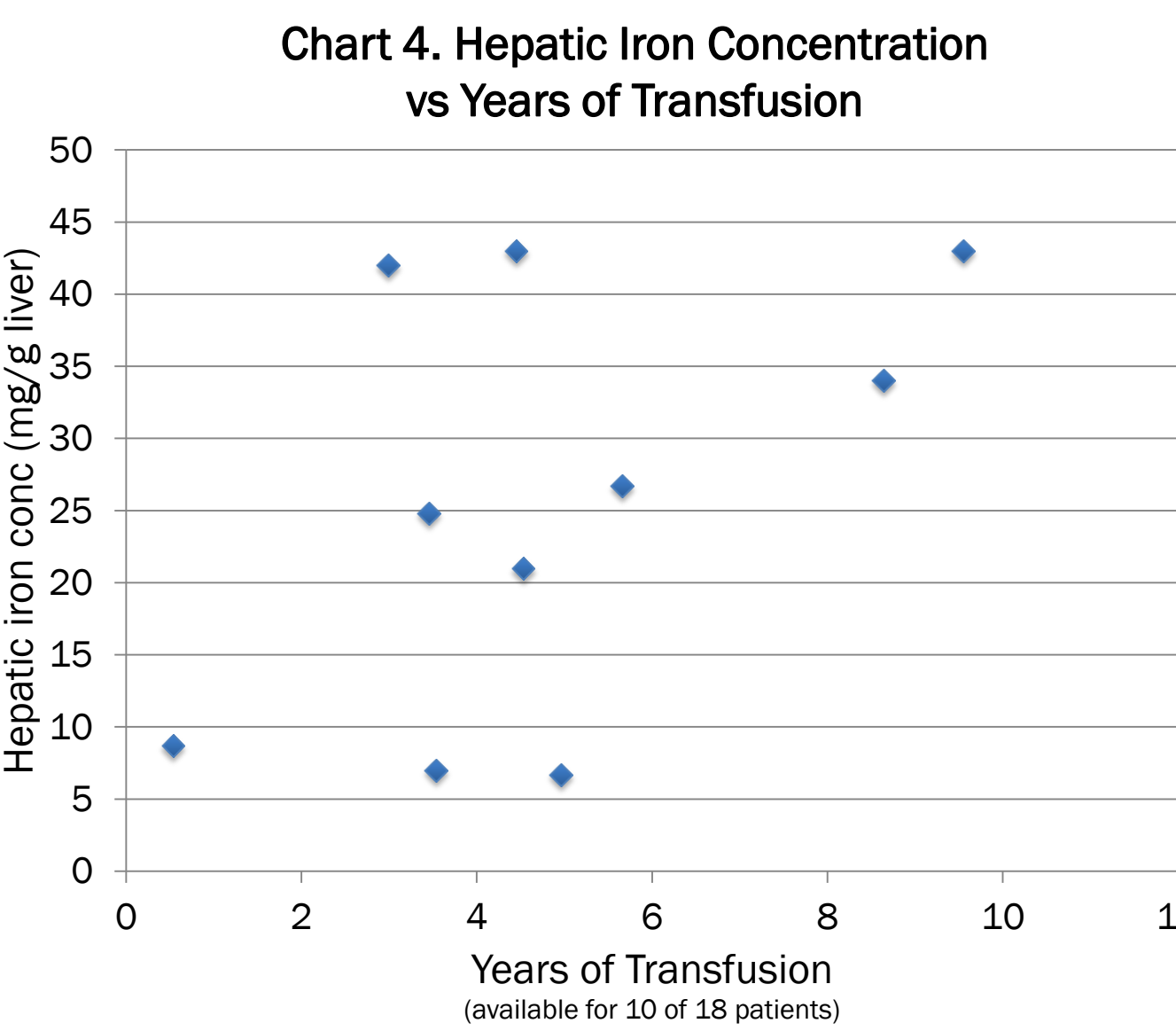
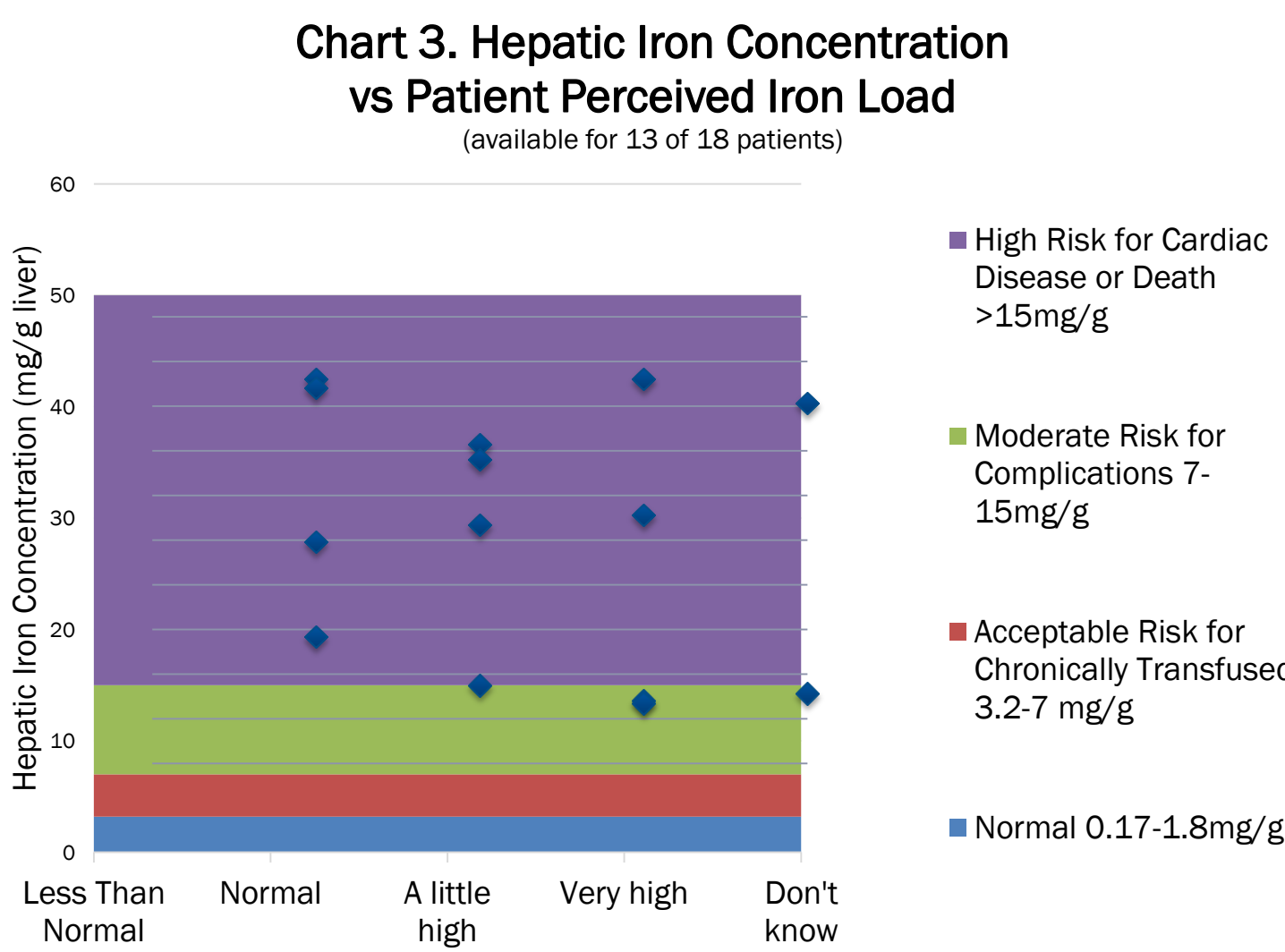


Table 2. Patient vs Caregiver Reported Barriers to Adherence				
	Subject Reported Barriers		Caregiver Reported Barriers	
	n=18	%N	n=18	%N
I never miss taking it	0	0%	-	-
Just forgot	14	78%*	8	44%
Not feeling well	5	28%	-	-
Interferes with activity	3	17%	-	-
Wasn't home	8	44%	-	-
Didn't get refill on time/ran out	7	39%	8	44%
Don't think I need it	1	6%	-	-
Hate the taste	13	72%*	-	-
Hard to swallow	3	17%	-	-
Side Effects	1	6%	0	0%
Worry about the long term side effects of Exjade	1	6%	-	-
I won't take it no matter what/ I refuse	5	28%	-	-
Exjade cost's too much/can't afford it	-	-	1	6%
Exjade is not helping my child	-	-	0	0%
I didn't understand the doctor's instructions	-	-	0	0%
My child does not cooperate with taking Exjade	-	-	8	44%
Other	-	-	6	33%
No Response	1	6%	1	6%

Table 3. Patient Reported Barriers to Adherence Stratified by Age						
	Child (5-9yo)		Preteen (10-12yo)		Adolescent (13-18yo)	
	n=6	N%	n=7	N%	n=5	N%
I never miss taking it	0	0%	0	0%	0	0%
Just forgot*	3	50%	7	100%*	4	80%*
Not feeling well	3	50%	2	29%	0	0%
Interferes with activity	3	50%	0	0%	0	0%
Wasn't home	1	17%	4	57%	3	60%
Didn't get refill on time/ran out	3	50%	4	57%	0	0%
Don't think I need it	0	0%	0	0%	1	20%
Hate the taste	4	67%*	3	43%	0	0%
Hard to swallow	1	17%	0	0%	2	40%
Side Effects	3	50%	0	0%	0	0%
Worry about the long term side effects of Exjade	0	0%	1	14%	0	0%
I won't take it no matter what/ I refuse	2	33%	1	14%	2	40%

Table 4. Patient and Caregiver Recommendations to Improve Exjade Adherence		
Subject Recommendations	N=18	%N
Improve Flavor/Taste	7	39%
Talk to parents more about it	1	6%
Oral Suspension	1	6%
Pill Form	3	17%
Chart to keep track	1	6%
I don't know	1	6%
No Response**	4	22%
Caregiver Recommendations		
Improve Flavor/Taste	7	39%
Tell patient how serious it is to take it	1	6%
Reward System	1	6%
Make it less expensive	1	6%
I don't know	1	6%
No Response**	7	39%
Medication Administration		
Caregiver Administered	8	44%
Subject Administered	2	11%
Caregiver and Subject Together	7	39%
Other	1	6%
No Response	1	6%

Acknowledgements

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Selected References

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Conclusions & Clinical Implications

The most common barriers to Exjade adherence were forgetfulness, and an unpleasant taste. There was a difference in most common barriers across age groups:

- Top barrier for preteens and adolescents were forgetfulness and not being home.
- Top barrier for children was poor taste.
- Common barrier for children and preteens was not refilling the medication on time or running out.

Limitations

- Patients from only one study site were evaluated.
- Only patients who were engaged in the sickle cell clinic were included in the study.
- Measure of adherence was not a direct measure because we did not observe patients’ intake of medications; also pharmacy records were not evaluated.

Future Directions

- Expand study to include patients at CHOA at Egleston and Scottish Rite.
- Evaluate barriers of patients who do not follow up regularly in clinic.
- Compare patient and caregiver reported adherence to compliance with medication refills by evaluating pharmacy refill records in addition to lab values.
- Implement reminder systems to improve forgetfulness, the top barrier across all ages.
- Transition patients to potentially more tolerable Deferasirox granules or Jadenu™ tablets to take into consideration patient and caregiver recommendations to improve taste and creating a pill form of the iron chelator.
- Repeat study in the future to evaluate if barriers to adherence improve after transitioning patients to more palatable alternatives.