

The Life of an ARPKD Boy

William Garrott, age 15
Carolyn Garrott, PharmD

The Garrott Family

Matt
Carrie
William
Matthew

Picture May 2021



My Background

- Graduated with PharmD in 2005 from Ohio Northern University
- Started career at Walgreens
- Had no special training in nephrology or pediatrics
- Did help with jargon, logistics
- Struck a balance health care professional vs parent

The Prenatal Era

- Due May 7th, 2010
- OB appointment April 21st, at 38 weeks
- No amniotic fluid on bedside ultrasound
- 2 hour ultrasound with MFM
 - Bright, echogenic kidneys
 - Was asked if I had taken ACE-inhibitors
- Admission to hospital and induced on Wednesday afternoon
- Assumption was fetal kidney disease
- NICU and pediatric nephrology brought in



42 Hours Later

Friday, April 23rd, early afternoon

- Fetal bradycardia led to emergency C-Section
- William immediately taken to NICU
- Placed on ventilator and oscillator
 - Pulmonary hypoplasia (underdeveloped lungs)
 - pneumothorax
- Imaging studies done
 - Echogenic kidney
- Advice from the NICU attending: “Cherish the time you have with him.”
- Preliminary diagnosis of ARPKD



Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- <https://pkdcure.org/about-the-disease/arpkd/>
- Combination kidney disease and Congenital Hepatic Fibrosis
 - All have varying degree of kidney and hepatic disease
- Nearly 80% of children with ARPKD survive the newborn stage.
- About one-third of these children may need a transplant or dialysis within the first 10 years
- Just 20 years ago, only half the children with this disease lived to see their 10th birthday.
- Today, more than 90% of children who survive the newborn period reach their 20th birthday, and half of them still haven't needed dialysis or a transplant.

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

Prenatal ARPKD

Symptoms

- Diminished amniotic fluid levels
- Enlarged kidneys
- Lung underdevelopment
- Lung functioning issues

Post-Birth ARPKD

Symptoms

- Enlarged kidneys
- Breathing problems
- Excessive urine production
- Hypertension
- Growth problems
- Congenital hepatic fibrosis
- Enlarged spleen
- Low red and white blood cell count
- Low platelet counts

Genetics

- Autosomal recessive disease
 - Need 2 copies of affected gene
- Genetic testing done
 - Dad has PKHD1 variation
 - I have a “VOUS” at same location on gene

NICU Era

- Stabilized
- Started urinating
 - Almost had PD catheter placed
 - Saved by observant NICU nurse
- Spent 10 days in NICU proper
 - Removed his own chest tube
 - Started taking bottle
- 20 Days in Step Down Unit
 - Main focus learning how to eat
 - Similac PM 60/40
- Discharged home 5/20/2010

Infant Era

Now What?

- Same issues as everyone else PLUS
- Feeding issues
 - Volume issues due to large kidneys
 - Calories vs volume
 - Keeping potassium and phosphorus low
 - SPS
 - Renvela
- Hypertension
 - Blood pressure monitoring at home on an infant
- Navigating child care and back to work

Working Mother Era

Why keep working?

- Income
- Insurance
- Professional fulfillment
- Break from home routine / get out of the house
- Working 30 hour week
 - FMLA leave when necessary
- Overnight shifts
- Mom guilt

Surgery Era

- June 2010
 - G-Tube Placement
 - Overnight tube feeding plus boluses during day
 - Loved to pull those out
 - Learned how to replace ourselves
- August 2010
 - Single nephrectomy
 - Broviac catheter insertion
 - Hydrocele repair
 - Peritoneal Dialysis Catheter insertion
 - 14-day hospital stay to manage leaking
- November 2010
 - Hydrocele / hernia re-repair

Pre-Dialysis Era

December 2010

- Admitted with fever on December 4th
 - Fever + PD catheter = automatic admission
- High fever, feeding problems, oxygen requirement
- Could not pinpoint source of infection
- Decision to remove remaining kidney
 - Will have to start dialysis
 - Required 2 surgeries to revise PD catheter
 - PD education
- Discharge home 1/5/2011



Peritoneal Dialysis Era

- 14 hours of treatment daily, plus daily dwell time
- Flowsheets with daily calls to PD nurse
 - Prescription changed nightly
- Weekly visits to PD clinic
- Twice weekly blood draws
 - IV access
- Daily care, weekly dressing changes
- Medicaid WAIVER program
 - Full time nursing care
- Hospital stay June 2011
 - High fever with sepsis
 - Klebsiella Oxytoca
- Pulled out his Broviac catheter at an appointment
 - Mediport placement

Peritoneal Dialysis Era, continued

- Developmental delays became more noticeable
- Traditional Day Care no longer an option
- Walked at 19 months, crawled later
- Help Me Grow services
- Occupational, speech, and physical therapy services
 - In home
- Feeding Disorders Clinic
 - Intensive feeding therapy

Hemodialysis Era

- PD site infections
- Remove PD cath, insert 2 neck catheters
- Four days per week, four hours per session
 - No dedicated pediatric HD; HD staff was NOT accustomed to infants
- IV antibiotics
- Ripped out one night
 - Emergency surgery to replace
 - Also put in new PD cath
- During this time, also reached the minimum weight required for listing on transplant list (12kg)
 - Immediately placed on, then off the active wait list



Pre-Transplant Era

- On deceased donor list
- Also worked up living donor
 - Living donor process
- Received several “almosts,” “back up to the back up” calls
- Primary nephrologist, Dr David Kenagy, to leave UH
 - Last day was Monday, April 30th at 5pm

Transplant Day

- Phone call Tuesday, May 1st, shortly after midnight
- Hospital at about 3am, last minute blood tests
 - Dialysis cut off early
- Transplant surgery at noon
- Surgery was several hours
- Admission to PICU
- 6-day hospital stay









Post-Transplant Era

- Eventual PD cath removal
- Gradual move to quarterly clinic visits
- Monthly lab draws
- Developmental Delays
- Help Me Grow
- LEAPS
- IEP

Post-Transplant Era

- Autism diagnosis
 - Related to kidney disease?
- Time to grow family
 - William finally stable
 - Chance to live “normally”
 - IVF with PGD
 - CVS testing

Specialists

- Nephrology
- Gastroenterology (Hepatology)
 - Congenital Hepatic Fibrosis
 - Semi-annual ultrasounds, fibroscan
 - Annual endoscopy
- Developmental
 - Autism, ADHD, anxiety
 - Neuropsych testing
- Surgery
 - 17
- ENT
 - Tubes, adenoids, tonsils
- Psychology
- Cardiology
 - Dilated aortic root
 - Annual echo

Part 2

William Garrott, Kidney Transplant Recipient
Age 15, Transplant 5.1.12
From Avon Lake, Ohio

I Do Not Remember Anything Pre-Transplant

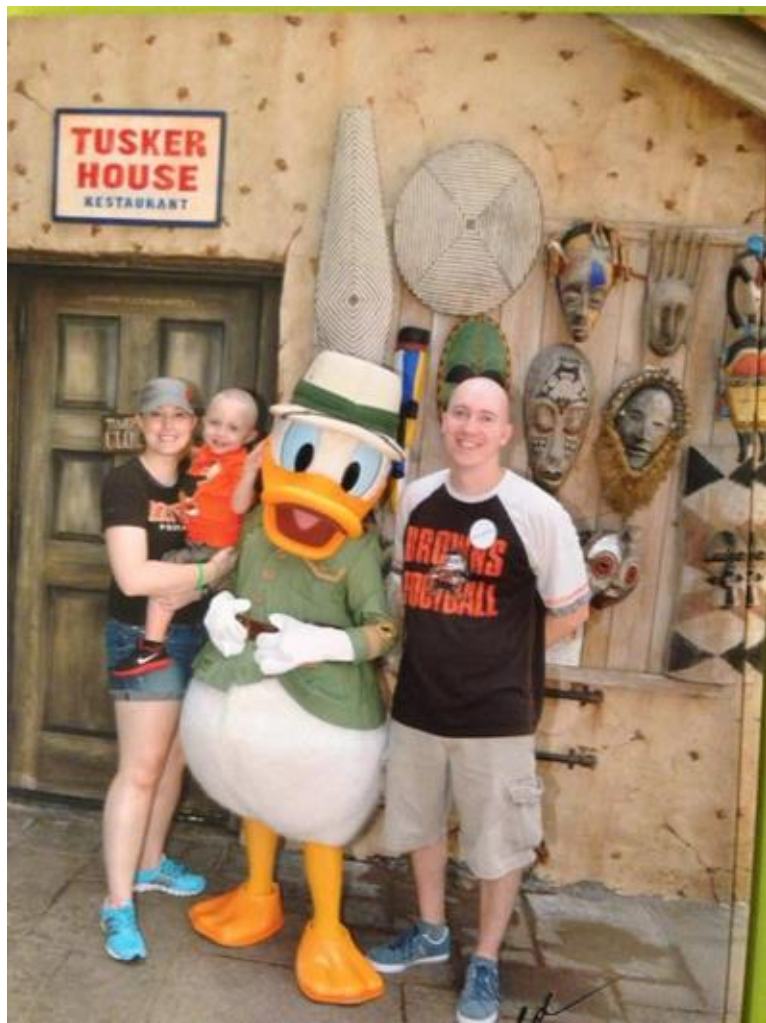
- But I do remember a few things:
- Watching movies while on Dialysis
- Nene watching me every day.
- Only ONE Hospital stay, I remember that it was in 2011.

Ages 2-5 Era

Transplant: I remember opening my eyes when I was being wheeled back for the big surgery, and then I went back to sleep, and didn't wake up until it was all over.

- Make-A-Wish trip in 2013
 - Disney World
 - Universal Studios
 - Give Kids the World Village





Age 2-5 Era, continued

- Ear tube surgeries
 - Lifelong fear of the anaesthesia mask
- Started LEAPS preschool
 - Local school district preschool program
 - Start of IEP
- My at-home nurse Nene
- Started getting in-lab blood draws done
 - EMLA cream
- Had 2 cats and one dog
 - Ben, Jerry, and Reese

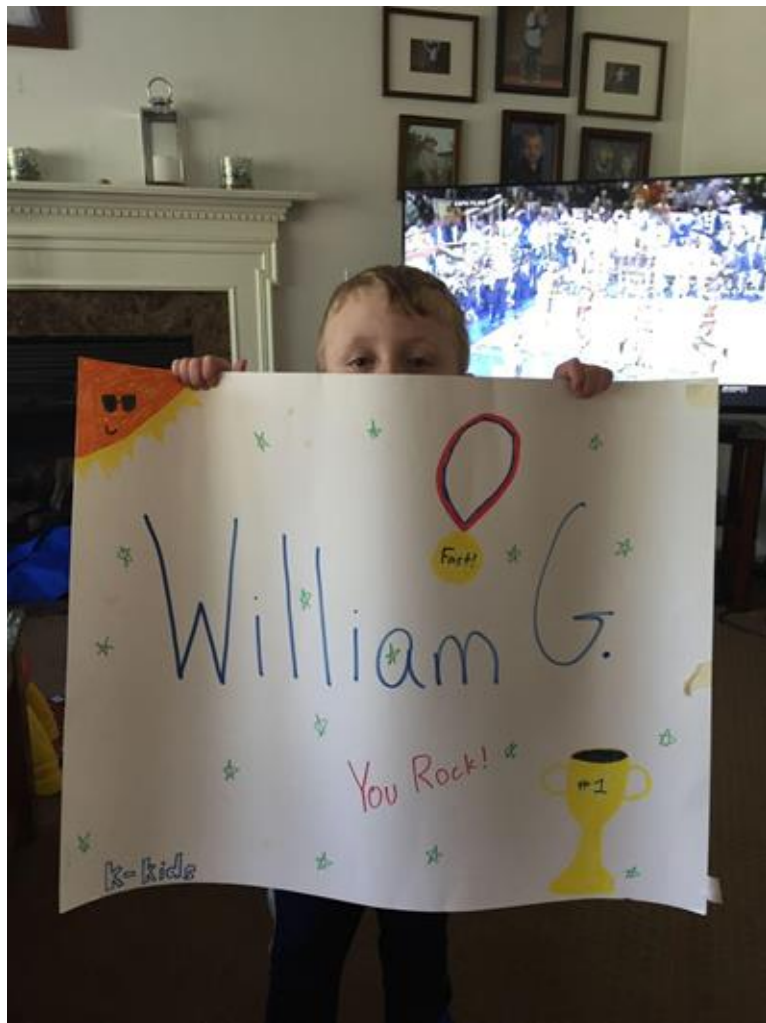




Elementary School Era

- Started Westview Elementary in 2015
 - On IEP, not attending “neighborhood school”
- No major restrictions
 - Unlimited access to water and restrooms
 - Regular phys ed, recess, just no contact sports
- Spent 80-90% of school day in normal classroom
- Rest of time spent with intervention specialists
 - Mrs Harlan and Mrs Slivinski
 - Mrs Patrella
 - Speech and occupational therapies as well
- Special Olympics
 - Track and field day
- Transition off of IEP and into “gen pop”





Elementary School Era, continued

- Dances
- Activity Nights
- Awards
- Soccer
- Kids on the Move
- Young Rembrandts



Westview
Golden Paw Award



For: being a good example
after nites waiting for
classmates to finish.

Given By: Baker

Classroom Teacher: Baker

Westview
Golden Paw Award



PKD Walk Era

- Annual walks to raise money for PKD Foundation
- Have raised thousands of dollars for the foundation



The COVID Era

- End of fourth grade into Middle School
- Sent to ER at the end of March 2020 with high fever
 - Negative COVID test and discharge home
- Remote learning
- Transition to middle school was less than ideal

Middle School Era Era

- Started playing drums / percussion
- Taking Spanish
- Jazz band
- Washington DC Trip
- Marching Band Helper

Travel

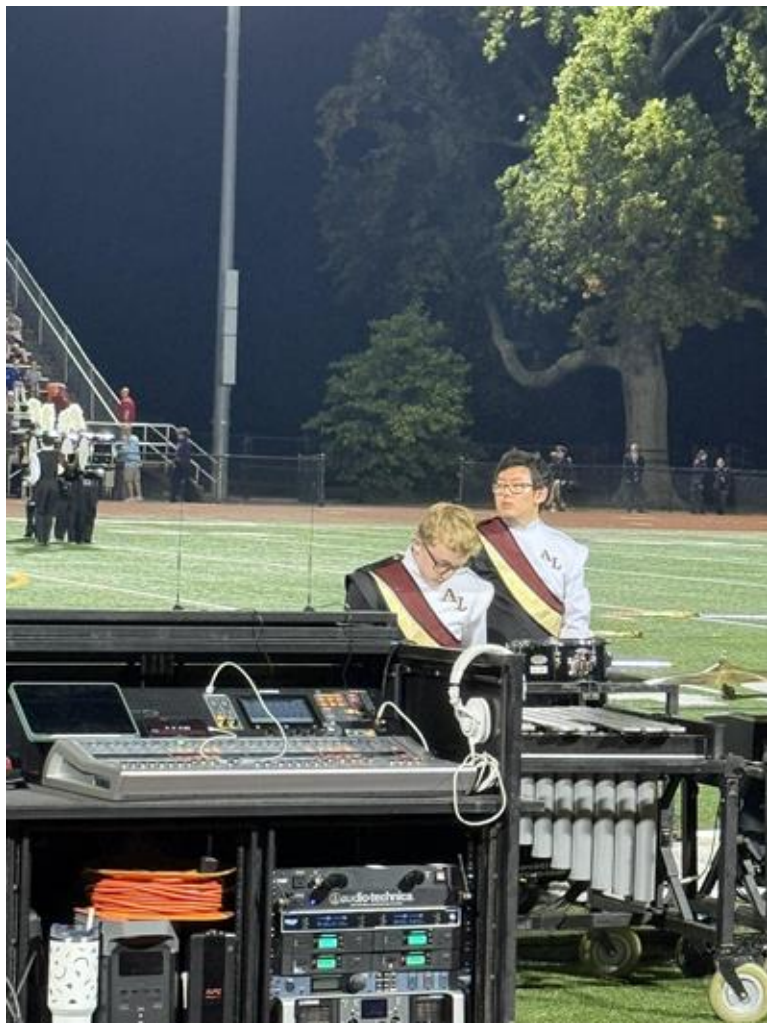
- Disney World
- Maryland
- Florida
- Deep Creek, Md
- Kansas

High School Era

- 504 plan for medical needs
 - Access to water
 - Restroom breaks
 - Other accommodations specifically tailored to autism / ADHD
- Kidney disease not detrimental to learning
- Can't participate in contact sports
- Otherwise a normal HS experience

Marching Band Era







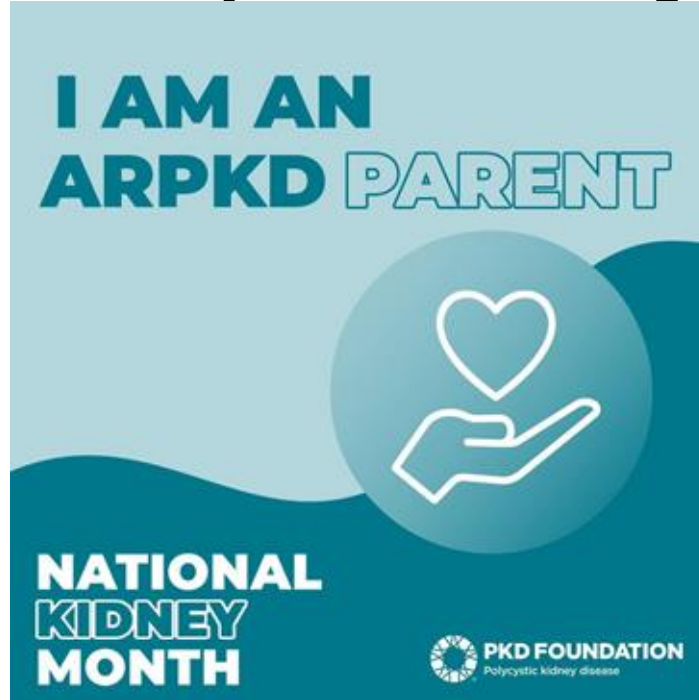
Daily, monthly, quarterly routines

- Daily water intake
- Meds three times daily
- Daily exercise
- Monthly lab draws
- Nephrology every 3-4 months
- GI every 6 months
 - Ultrasounds and endoscopy yearly
- Cardiology with echo annually
- Yearly vaccinations
- Importance of developing good relationships with care team

Current Med List, November 2025

- Prograf 4mg BID
- Azathioprine 75mg QD
- Propranolol 10mg BID
- Amlodipine 7.5mg QD
- Ursodiol 125mg TID
- Escitalopram 10mg QD
- Ferrex 150mg QD
- Vitamin C 500mg QD
- Metadate 20mg QD
- Probiotic QD
- Magnesium 500mg QD
- Multivitamin with Vitamin D QD

Thank you for listening!



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Time for Q&A



Links and contact info

<https://my.clevelandclinic.org/patient-stories/627-pediatric-kidney-transplant-patient-is-thriving-10-years-later>

https://www.nxtbook.com/nxtbooks/pkdlife/pkdlife_2022summer/index.php

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