

INTO THE UNKNOWN



Living with Children's
Interstitial Lung
Disease

- Our Journey with Sarah

In the beginning



- Sarah born at 34 weeks gestation uncomplicated natural birth. 3rd child.
- Airlifted to RHW with acute respiratory distress.
- Intubated. On HFV and NO₂ for 3 weeks, normal ventilation 2 weeks, CPAP ~ 1 week, weaned to nasal cannula O₂. XRays & blood tests – too many to count
- Lung biopsy @ 3 weeks old to test for rare lung diseases - results inconclusive
- Blood tests for common surfactant protein deficiencies - negative
- Cystic fibrosis tests - negative.
- Blood samples sent to Larry Noguee at JHH for more complex genetic testing.
- 5 doses of artificial surfactant, multiple doses of steroids and antibiotics administered.
- Transferred to Wollongong Hospital NICU after 2 months at RHW
- Sent home after 1 month on oxygen 0.03L O₂. NG tube for comp feeds.

Surviving the first 2 years



- Breastfed till 14 months. O2 saturation check revealed sats avg. 85%, oxygen - increased to 0.25L/min where it stayed till she was ~18 months old.
- After swine flu (July 2009) O2 requirements increased and have stayed at 0.5-0.75L ever since. Pulse oximeter used for spot checks regularly throughout the night.
- Review of lung biopsy a final diagnosis of ILD was given, with a best fit complex diagnosis of pulmonary interstitial glycogenosis (PIG) in 2009.
- About 20 hospital admissions. Memorable admissions for swine flu, pneumonia and parainfluenza 3 (went home on CPAP). Vomiting a major issue. Losec trial. Now Nexium. Hydroxychloroquine trialled for 3months.
- FTT - Has finally made it onto 5th percentile for weight Jan 2010. Now fed on 1.2 cal PediaSure with fibre plus Calogen supplement. Weaning onto 1.5 cal Nutrini Energy Multi Fibre.
- Constant pulling out of her NG tube and FTT meant that an AMT Mini-One gastrostomy button was inserted Feb 2009. Decision made to not do fundoplication. Best thing we ever did.
- Oral feeding is still poor - PEG for the majority of her calorie intake. She has 3 bolus feeds of 100mL during the day and 400mL continuous feeding overnight. Drinking/swallowing skills improving.
- Synagis (RSV) vaccination approved during winter months (has not had RSV yet ☺)
- Gross motor skill delay but fine motor, language and cognitive skills excellent. Finally walking at 2 years old.

The S-Team

- Paediatrician - Phil Goodhew ✓✓
- Respiratory Physician - Adam Jaffe
- Sleep Specialist/CPAP - Arthur Teng
- Paediatric Dentist - Hayfa Hadi
- Opthamologist - Rajiv Shah
- Orthoptist - Diana Taylor
- Paediatric Surgeon - Bruce Currie
- Gastrostomy CNC - Allison Kennedy
- Physiotherapist - Samantha Stephens (Wgong Hosp)
- Dietician - Jane Cleary (Wgong Hosp)
- Speech Therapist - Anna Wheaton (Wgong Hosp)
- Cardiologist – Christoph Camphausen
- GP – Cathy Allen (care plan being created)



A Ward at home

- Pulse Oximeter
- Graseby Monitor
- Kangaroo Pump
- A good pair of ears tuned to her breathing rate 😊
- Resmed CPAP (30cmH₂O, pressure 7.0)
- Oxygen Concentrator (3-4L/min via CPAP)
- 2 large E + 15 CH O₂ cylinders
- New family van to transport equipment & kids
- Air conditioning
- Air Purifier / Ioniser
- Medical consumables – you name it, we have it
- Formulas and Medication



Full-time Job

- Mum is –
 - Full time Carer
 - Full time Mummy to three kids under 6
 - Full time PA for Sarah
 - Part time Environmental Chemist
 - Wife when I get the Chance
- Dad is –
 - Full time Daddy
 - Medicine pickup man
 - Oxygen pickup man
 - Full time IT Manager
 - Husband when Wife gets the chance ☺
- There has barely been a week in Sarah's life that we have not attended some sort of medical appointment or been in hospital
- At least 2-3 hrs spent daily attending to Sarah's individual needs - cleaning feeding equipment, changing oxygen cylinders, making phone calls, organising supplies/medications
- Hard to make time for Neve & Noah – restricted to where we can take Sarah and hefting O2 cylinder is a pain in the ***



Strapped for Cash



- Private health insurance only covers so much.
- \$18000 out of pocket in first year
- \$220 per week for capable Nanny
- Medication not on PBS e.g. Anti-reflux
- Electricity bill increase (air con, concentrator)
- Parking fees/petrol/food when in hospital
- Retail therapy
- Medical appointments every week

Imagine...

- Wondering if your baby will live and for how long
- Living apart from your 2 other children for six months
- Being isolated from most of the community for fear the GERMS MIGHT GET HER
- Your 5 year old apologising for getting sick and making Sarah sick
- Your kids being expert vomit catchers
- Your mobile child being tethered to an oxygen cylinder 24 hrs/day



The System



- The Good
 - Direct admission to Childrens Ward, bypassing Casualty
 - In general the medical and Allied Health staff now know Sarah very well, respect our opinions and work with us in caring for her
 - Sarah likes hospital food more than home food!!!!
- The Bad
 - The medical record system – we still have to give her medical background every admission.
 - Risk of further infection in shared rooms
 - Not enough communication between medical providers and parents at times
 - Childrens Ward not adequately equipped/prepared (in our case for CPAP, low flow regs.)
 - Final biopsy/genetic testing results delayed (months)
- The Ugly
 - Uninformed medical staff not listening to informed parents = late NETS intervention and intubation that could have been avoided.
 - Living in the Nurses Quarters (Coulter House) ☹️ now condemned
 - Big hassles with changeover from Resp. Health to Enable (Sorry you haven't put your order in by the cutoff time - your baby will have no oxygen for the weekend)
 - NG tube for 12 months – was she lost in the system???

Who ya gonna Call?



- Other families with similar problems (we've found 2) ✓✓
- Govt Crisis Payment available up to \$10000
- Govt Carers allowance
- Barnados – volunteer support weekly, help with bill payments, gifts and entertainment for siblings
- Early Childhood Clinic – weekly home visits
- chILD Foundation - www.childfoundation.us
- Miracle Babies Foundation – creation of Illawarra Playgroup
- Allied Health Services (SESAHS) –Premmie Clinic
- Disability Trust (Kids Boost Funding) – nappies and formula
- **Enable – 02 and PADP equipment**

Saving our sanity



- Science/medical backgrounds, living close to Wollongong Hospital, previous NICU experiences and living with close family in Randwick helped us deal with the overwhelming situation early on.
- Medical staff always recognise our interest and understanding and take the time to clearly and truthfully explain things to us. We are thankful that Sarah is our 3rd child, meaning we only have sick child worries and not 1st baby worries as well.
- It has been an emotionally and psychologically draining time for us all, and the stress of caring for a chronically ill child has affected us all. Darren and I are both on anti-anxiety medication. Our greatest comfort has been speaking with parents of similarly ill children. The chILD foundation website has been a great source of information and comfort for us.
- Our time in hospital with Sarah has taught us so much and we are all better people for the experience. We are kinder, more patient, resilient and organised than we ever were and have grasped the true meaning of “don’t sweat the small stuff”.
- Co-operative baby. Supportive network of family and friends. Understanding workplaces.
- Complex patients such as Sarah need a case-worker to help manage things. Caring for 3 children has left Darren and I with zero time for ourselves. Things are improving slowly but we find it difficult to leave Sarah with anyone because of her special needs. Generally we have been left to our own devices to source information.

A new day

- New alveoli until age of 4 – growth is critical (finally on the growth charts for weight)
- Reduced occurrence of URTI (CPAP and Azithromycin helping)
- Prednisone working when acutely ill (Longer term weaned dose works better than pulse treatment)
- Longer periods out of hospital = happier family life
- Now walking and talking (LOUDLY)
- Address feeding issues
- Organising Pre-school + Aid for 2011
- Oxygen weaning ??
- Think about lung transplant ??? Hope not



Thankyou for Listening

