



# Parley

Parents And Relatives  
Listening and Empathising  
with You

## Hayley's Story

### **CCAM, BPS & NYSTAGMUS**

(Congenital Cystic Adenomatoid Malformation )(Broncho Pulmonary Sequestration)

It was discovered at my 20 week anomaly scan that there was a problem with our unborn child's lungs. The sonographer kept going over the same dark areas on screen which I new meant fluid, I thought it was the kidneys but she said no it's babies' lungs and then switched off the screen.

I was in tears my husband was trying to reassure me but not knowing what was wrong was just a horrible feeling for us both. We then spoke to a midwife as consultant had gone home who really didn't no herself what the problem was just that there were fluid filled structures on both lungs.

That's what was so hard still, the not knowing, then the midwife arranged for us to go up to St Georges Hospital in London for a scan and consultation the very next day.

As soon as we got home I was straight on the internet trying to self diagnose and the only thing that came up was CCAM (Congenital Cystic Adenomatoid Malformation) never heard of it ,all the web sites were scaring me, some said it was best to terminate, others said about operations on the unborn foetus, still birth was common... that was enough I had convinced myself that this is what my baby had and it was going to die.

The journey up to St Georges was silent, seemed to take forever, then the next thing I'm in a high tech scan room with loads of people watching my scan then the consultant said he agreed with the Worthing sonographers findings and explained that it's not exactly understood why it happens but it's nothing I had done wrong or no abnormal genes from either of us which we were both thinking at the time. He said that at some point usually around 6 weeks gestation the lungs form bronchials and with our babies the tubes had breaks in

them and in turn fluid filled cysts had formed in place, I asked what was this conditions name he said CCAM .

Our faces dropped as we were taken to a side room to be explained more, we were told it's a very rare lung condition what makes it even more rare is that it is in both lungs and only usually in one.

I then asked if they had seen the sex he said it's a girl! I couldn't stop crying we had been waiting for a little girl for 9 years we already had 2 boys and the thought she may be taken from us was devastating,

The consultant was very reassuring and said we will do every thing we can for her, the danger was of the cysts growing and displacing the heart which had started to happen already, if they grow too large other organs can be affected and something called hydrops can happen in which will be apparent through the mother showing signs of preclampsia. If this were to happen baby needs to be born or if too early draining fluid from the cysts in the unborn foetus was possible.

It was all so much to take in we went home like zombies then had to repeat it all to family and friends

I thought I would never stop crying.

I was told I would have to be very closely monitored every week at Worthing for check up and scan then every 2 weeks up at St Georges for a scan, had to have 2 sets of maternity notes as still were not sure where I would be delivered.

I took it upon myself to find out as much information as possible tried to find other families but only 1other CCAM baby had been diagnosed at Worthing in the past 10 years and couldn't find them, but thanks to the hospital of Cincinatti in the US they had a fantastic informative website with success stories and videos which was much needed, i now new more about it than the consultant at Worthing!

The feeling of despair started to vanish with every scan as the cysts were staying the same size, with encouraging words from the doctors.

It was my last scan at St Georges 2 days before Christmas and it was good news the cysts had not grown too much on the right and were hardly visible on the left, as a treat the consultant gave us a free 4-d scan so I could see my little angel, she looked perfect !

We were then introduced to our daughter's surgeon; he explained that she would need an operation at some point maybe in the first few weeks maybe not till she's 3 years old, but the cystic part of the lung was non functioning and could become cancerous in later life, that was the first mention of cancer which made us just want those things out of her as soon as.

I was given a date for delivery at St Georges for Feb 22nd 2007 1 week early as the longer inside the better for her lungs. We were shown the NICU and shown the breathing machine she may have to go

on and they explained she would be taken straight away from me to be checked but my husband could stay with her which was upsetting but obviously I understood the need.

22nd of Feb. arrived and after a long induced labour little Clio-Belle was born screaming weighing 7lb 8oz she looked just perfect how could there be anything wrong? She was breathing for herself and once she had been thoroughly checked I was allowed to hold her and give her a breast feed before being taken off to NICU, I was taken to my own room for rest as late at night but all i wanted to do is be with my daughter ! My husband gave her bottle feeds through the night and i fed her in the morning, she was doing so well just had this feeling all was well and couldn't wait to take her home.

We arrived home 2 days later with an appointment for a ct scan at 3 weeks old.

Three weeks past and the scan had been done now we had to wait another 2 weeks for results we were also given an appointment to see a lung specialist as well. Clio-Belle was such a content baby she had given us so much worry but already so much joy.

The results were in and as expected the cysts were there in the right lower lobe of her lung but miraculously they had disappeared in the left which has been known but not commonly seen we were chuffed to bits , but then we were told something else had been picked up Clio had an abnormal artery attached from her heart to one of the larger cysts feeding it with it's own blood

supply it was called BPS (Broncho Pulmonary Sequestration ) usually a condition by itself yet again another rare congenital condition plus the CCAM it made Clio more than 1 in a million !

We were told the BPS could mean that the abnormal artery could suffer an aneurism and be fatal for Clio as she got bigger and started to run around so it was decided then she would be operated on at 5 months to remove half her right lung and remove and repair the artery, it was at this appointment I noticed Clio's eyes had started moving side to side involuntary like she found it hard to focus so another appointment this time in Worthing for her eyes.

Clio-Belle had now been diagnosed with congenital Nystagmus, this just about broke me and Chris she had been through so much now waiting for her op to make her all better but Nystagmus was for life and no operation can fix it once again i was on the net looking it up and this time there was lots of information of yet her third rare

complication!! but it was another wait and see thing were in some kids the wobble of the eye gets worse sometimes it stabilises but i had to be strong for Clio and be positive there were options to help like glasses, when older extra large text on books extra time to complete exams, so we just put it to the back of our minds until op was over although hard as it was so noticeable to us didn't want people to stare but everyone was great about it so we agreed not to dwell and wait till her Ophthalmologist appointment to reassess at age 1.

Clio-Belle was 5 months old now and being prepped for surgery we were holding it together until Chris my husband took her down to theatre were we both broke down all our emotions came out then and we hugged and cried and let it all out. Then the waiting game began. After 4 hours in surgery we were taken down to PICU (Pediatric Intensive Care Unit ) and saw our little princess with wires everywhere and a large drainage tube in her back the tears came again but we were so pleased to see her, the surgeon said all went well but he also had to do a diaphragm repair as the cystic lesions had pushed into it, but all was well just recovery time now she was so out of it on morphine the first day bless her but by day 2 she was back to her smiling self all the nurses fell in love with her as she was so happy all the time and they were so fantastic with her you soon forgot about all the medical equipment.

After 3 days she came out of intensive care and back on the normal ward after 1 night there the next day her drainage tube was removed and her wound re-dressed , we couldn't believe how neat it was already healed on the outside it was about 4 in long around the bra line .

Soon as we came home she was back to her playful self you would never no today what she's been through ! Her lung will carry on growing until the age of 8 so will always be smaller but more than what it is now.

As for her eyes Clio has 20/20 vision and her Nystagmus has calmed she is lucky in the fact her focal point is straight in front as some kids with Nystagmus it is to the side so have to tilt their heads to achieve their best focal position, so with clio you only really notice it when she looks up or to the side she has one of the milder cases which obviously we are glad she has had so much in her life already it did seem a little unfair! But we would have loved her just the same even if her Nystagmus was severe!!

Clio-Belle is 18 months old now and doing all that she should be doing for her age, the surgeons did a wonderful job and her scar is so faint you would have to look twice to see it, there is light at the end of every tunnel and we have now found it ,Clio will have 6 monthly checks just to keep an eye on her as her body went through so much but you wouldn't no it by looking at her and I wouldn't change her she is our 1 in a million !!!!

