

## Raine Family Story

My wife Natalie and I have five children, two of whom, Nicole and Lola, have ARPKD. Both have now been transplanted and are doing exceptionally well. Our story begins with the birth of our first child, Nicole.

My wife had a good pregnancy all the way through, with normal ultrasounds. We were both really excited about the birth. When Natalie was 38 weeks pregnant however she noticed that the baby was not moving so she went to the antenatal unit at our local hospital. An ultrasound scan showed that baby Nicole had very little fluid around her in the womb and that her kidneys were very enlarged.

Natalie was induced straight away and Nicole was born. It was a very worrying time as we didn't know what was wrong. Nicole stayed in the hospital for a week after the birth. There were no signs of any problems other than that her abdomen seemed distended but then we hadn't had a baby before so we didn't know if this was normal or not.

We attended an appointment at our local hospital and were told that Nicole likely had ARPKD. She was referred to a specialist paediatric nephrology unit but her appointment was several weeks away. At about 8 weeks old Nicole became very distressed and was crying a lot. Natalie noticed a lump in Nicole's groin and we quickly rang NHS Direct. We were told to take her straight to A&E. We took Nicole to our local hospital and she was transferred to the specialist centre by ambulance. She had a hernia as her tummy was so full of kidney and liver. Her blood pressure was very high indeed. Nicole had her hernia operation and was treated for both ARPKD and hypertension. That's where it all begins

Nicole was very ill as a baby. Her blood pressure was difficult to control and she was on an immense amount of medication. She spent a lot of her first year in hospital. She had several scans which found she had cysts everywhere. It was so scary. Nicole spent many years in and out of hospitals and being very ill at home too. As a baby, blood pressure control was the big issue. As she grew she suffered from cholangitis of the liver which made her very ill.

After meeting with the consultant's at the specialist centre it was clear that Nicole needed a combined liver-kidney transplant. This was a scary prospect as the risks were far higher than with a kidney transplant alone. However Nicole kept getting so poorly and spent so much time in hospital that we realised this was the right way to go.

On the 7<sup>th</sup> April 2009 at the age of 7 Nicole was called in for the transplant. It was surreal - we were happy but also so nervous. Nicole went to theatre the following morning. She was in there for about 10 hours. Everything went well. When we saw her in ITU afterwards she looked so different. She was all swollen and the machines were terrifying. We wanted her to cry or complain but she didn't. After just under two days in ITU, Nicole was moved onto the ward on high dependency. Things were looking good. After that she sailed through recovery with no problems and was back home after just 14 days. She hasn't looked back since. A healthy child at last, able to do everything other kids can.

Lola is our 4<sup>th</sup> child. This time was different. Natalie had regular scans as well as regular heart beat and movement monitoring. Lola was diagnosed with ARPKD at the 12 week scan. Natalie was induced at 38 weeks and Lola stayed in hospital for a week after her birth. She had high blood pressure but it was easier to control than Nicole's and Lola spent far less time in hospital. Lola had a hernia operation and more dietary restrictions than her sister but pretty much followed Nicole's trend in how her kidney function deteriorated. She was also put on the transplant list.

Lola was called in for transplant one Friday. We were anxious. Lola was younger than Nicole was when she had her transplant. Although we knew what to expect this time, we also knew that it would not necessarily go as smoothly as it did with Nicole. It was nerve racking but in the end we

got sent back home as the new organ was not suitable - it was too big for little Lola. We got home with a little disappointment but also a big 'phew'.

We thought it would be a while until she would be called in again but the following Saturday night the call came and off we went. We arrived fully expecting it to all go through this time but again the organ was not suitable and we went home to await the next call. Then about 2 months afterwards on the 19<sup>th</sup> of November 2012 Lola was called in for the third time and this time it went ahead. She was transplanted the next day and was in theatre for pretty much the same amount of time as Nicole. Lola's recovery was different to Nicole's and there were a few more worries but after about 16 days Lola was discharged back home. Now both girls are living normal lives and enjoying good health.

The difference in the girls is amazing. The energy they have and their happiness. They smile and laugh so much more and I'm quite sure that a bit of cheek and attitude was also transplanted! But we don't mind. They have healthy, fun filled lives ahead of them now. Huge thanks to our consultant paediatric nephrologist, hepatologist and the entire transplant team.