

Incontinentia Pigmenti - small, but not insignificant

by Karen Mosely

My daughter, Katie, is 11 - she arrived 4 weeks prematurely, was footling breech presentation and had to be delivered by emergency caesarean section - a bit of a shock to all of us, including Katie!

Whilst in hospital for the first week I had difficulty feeding her; the midwives said she was a very 'windy' baby. Just prior to discharge a junior doctor saw her being 'windy' and promptly rushed her to the Special Care Baby Unit - the 'wind' was her having convulsions. This was all a bit much to take - my little girl in an incubator covered in wires and on phenobarbitone.

However, the drugs stopped the convulsions* and we thought we'd be going home; but then the 'nappy rash' that had been virtually ignored spread up her trunk and over her arms and legs - What now?! *It's thought that the fits were caused by her prematurity/delivery - not connected to IP.

Luckily for us, as I now know people have had to wait months or years for diagnosis, the attending Dermatologist thought immediately that it could be Incontinentia Pigmenti - asked for a biopsy and confirmed it. Although I said luckily - at the time we didn't feel at all lucky. Nothing seemed to be known about this genetic condition. The paediatrician (a lovely man) went from books and could do no more than reassure us that the fits had stopped for now and tell us that Katie would be carefully monitored. We got worried about all the things that might happen later - he didn't know - nobody did.

She was carefully monitored, measured and assessed - but apart from a slightly larger head than normal (later dismissed as a family trait!) and a tendency for clumsiness everything seemed OK. The rash got worse, then cleared occasionally coming back if she was ill, but most things were just like all the other babies we knew.

The reason for her clumsiness was later found to be that she had no sight in her left eye (this hadn't been a clear possibility in any of the literature that we had at the time) and was one of the biggest shocks since the fits. Katie, though, didn't know any different - they think she's never been able to see from it, and got on with life very happily.

I didn't know anyone else with IP until, when Katie was about 5, I heard about Contact a Family through my Mum who was a health visitor at the time. I got in touch with them who put me touch with a couple of people (the ED group being one of them).

Contact a Family have since given my number to a few people and we have had lovely chats and shared our experiences. It was so nice to know that we weren't the only ones - how I wished that these people had been contactable when I had Katie.

As it has turned out, apart from her eye, IP doesn't affect Katie too much - she's just started Grammar School and enjoys life to the full. But if I'd had someone to talk to at the beginning, I'm sure I would have had far fewer sleepless nights.

IP, in general, isn't particularly debilitating - but not knowing what lies ahead and feeling alone can be very debilitating for the parents.

Over the years I have spoken to a lot of people for long periods of time and then never heard from them again - which is great; knowing you're not alone and that someone is at the end of a phone if necessary is often all that's needed.

Having a link up for parents is also important so that the professionals have a point of contact for future parents, and so that any new research developments can be shared.

We may be a small group, but we must not be insignificant!

Some names and locations have been changed in order to preserve confidentiality. This article was first published in our newsletter (Volume 2 Issue 2 - January 2002)

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