

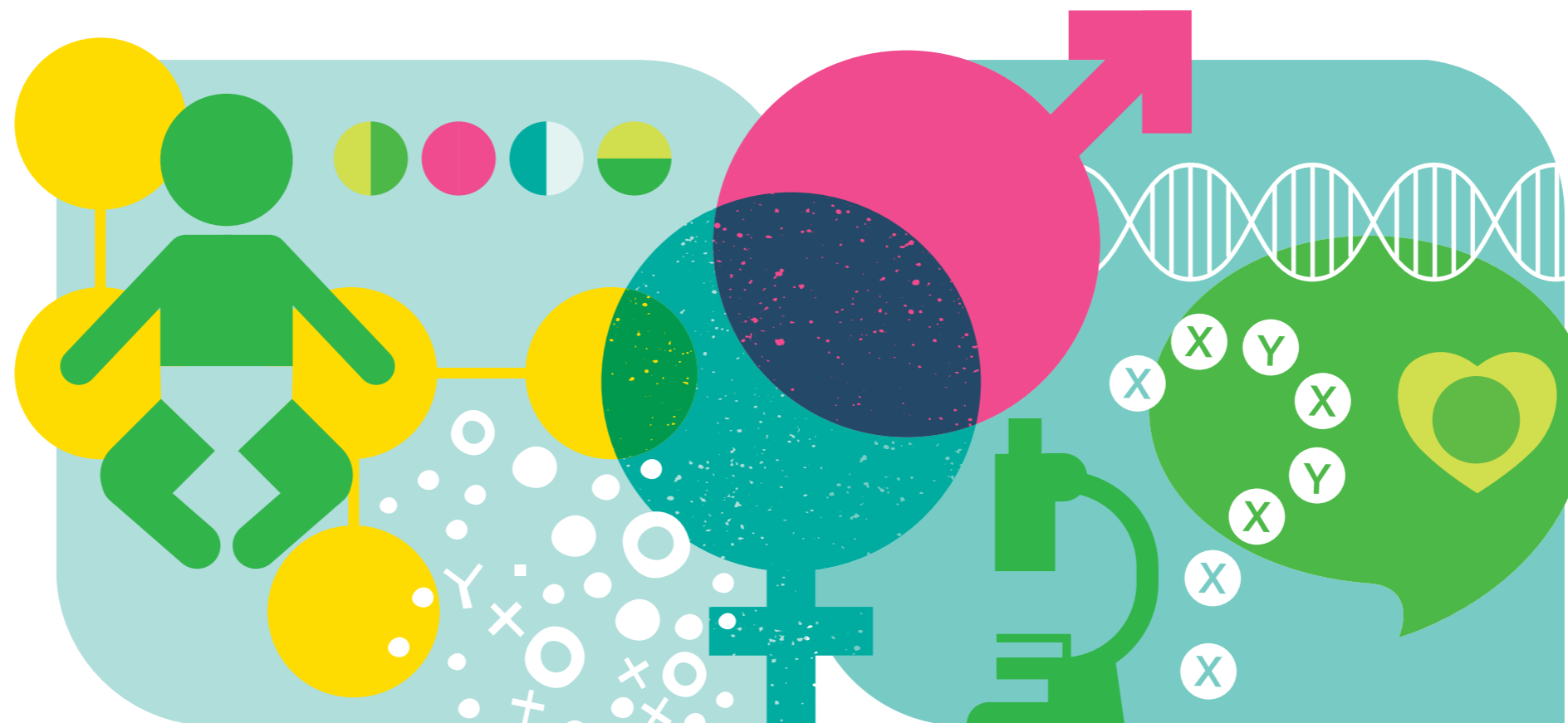
IS IT A BOY OR A GIRL? THIS IS OFTEN THE FIRST question asked by and of new parents. But what if the answer is not obvious?

Exact figures are unknown, however, it is estimated that around 130 to 140 babies are born in the UK each year with differences or disorders of sex development (DSD) (Personal communication, 2015) and, for most new parents, it will be an unexpected piece of news. For many, it will be the first time they have even heard of DSD and it will almost certainly be a time of extreme stress and worry until a multidisciplinary team can establish whether a baby is best raised as a boy or a girl.

It can be difficult for midwives too, yet they are in a position to reassure and normalise, as much as possible, in those early days of uncertainty.

A brochure has been put together to help families – and indeed health professionals who are not specialists in DSD – face these challenges. *When your baby is born with genitals that look different – the first days* provides key information to equip and support midwives and others to deliver the most thoughtful care at a difficult time. The booklet, which is available in print and online, has been developed by specialists in the field, such as medics and psychologists, as well as parents who have been through it themselves.

John Achermann, professor of paediatric endocrinology at the University College London Institute of Child Health and coordinator of the DSD multidisciplinary team at Great Ormond Street Hospital (GOSH), is one contributor. He explains that DSD is a blanket term for a lot of



Julie Griffiths explains disorders of sex development and what midwives can do to help parents understand their child's condition.

conditions that can present at different stages in life.

'Most common is when a baby is born with atypical genitalia, though we are increasingly getting families referred prenatally,' he says.

John adds that sex development is complex, with the sex of the baby not being determined by one thing. He explains that there are three broad categories of DSD.

The first is in a child with XY chromosomes whose testes do not form as expected or do not produce typical levels of testosterone. Or it may be that the body has not responded to the testosterone signals as expected. This means that the genitalia may look more like a girl's or somewhere between a typical boy and a typical girl. The second category is when a child's chromosomes are XX and the baby has higher levels of testosterone than expected resulting in an enlarged clitoris. Most commonly this occurs from the adrenal gland and this condition is called congenital adrenal hyperplasia (CAH).

The third category is where, instead of every cell in the body being XY or XX, there is a mix; some cells have a Y chromosome and some may not.

It can be important to understand which

condition a baby has when deciding whether the child is best raised as a girl or boy and, to do that, various tests are run. John says that the uncertainty is difficult for parents: 'There is often a fog initially, with lots of information to take in, but usually, after a few days, a clearer path emerges.'

GOSH clinical psychologist Polly Carmichael says that, when it happens, DSD can feel like a traumatic event to new parents. They have many fears for the present and the future, such as how their child might develop, what will happen at puberty and the implications for the newborn as an adult. Midwives can help manage the uncertainty, says Polly, though she acknowledges that DSD can be a challenging issue to face. Families may seek information from midwives that is not available and perhaps ask for an opinion on what the gender is likely to be. Polly says it is important to avoid guessing.

'Often, healthcare professionals want to make things better and give solutions,' she says. 'But it is unhelpful to second guess in those early days because if it is a wrong guess, it creates problems. Instead say things like: "It seems

DSD STATISTICS

130-140

- It is estimated that there are around 130 to 140 babies born in the UK each year with DSD. (Personal communication, 2015)

300

- Hypospadias (when the opening of the penis is not at the tip but along the shaft) is more common with prevalence one in 300 boys' births. (Djakovic et al, 2008)

13%

- It is estimated that 13% of infants with hypospadias and cryptorchidism (the absence of one or both testes from the scrotum) will have a chromosomal anomaly. (Society for Endocrinology, 2011)

4500

- Genital anomalies are estimated to occur in one in 4500 live births. (Calleja-Agius et al, 2012)

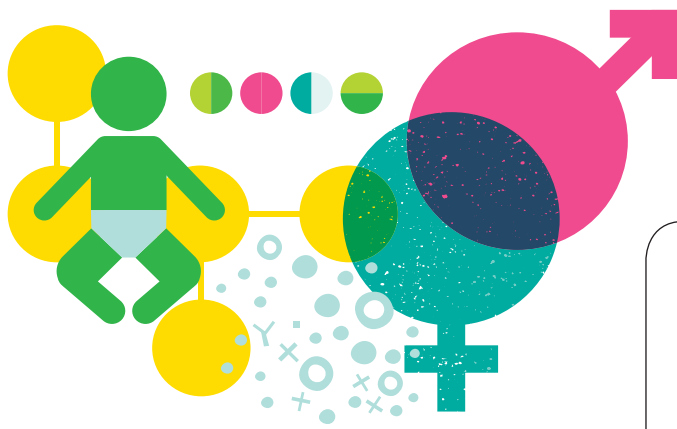
18,000

- Around one in 18,000 children have CAH. Most girls with this condition present in the newborn period with atypical genitalia and are at risk of a salt-losing crisis. (Khalid et al, 2012)

42

- The number of days parents have to register their baby's birth is 42 days in England, Wales and Northern Ireland. In Scotland, it is 21 days. This deadline can be extended in exceptional circumstances though this is rarely necessary.

Dealing with DSD



→ uncertain but we will get an answer. We'll do that as quickly as possible and, until then, we need to keep an open mind".'

Ellie Magritte believes that midwives have an important role to play in empowering new parents dealing with DSD. Thirteen years ago, she gave birth to a baby girl by emergency CS and, after she and her husband shared the good news with family and friends, they learned that her little girl's swollen labia may have testes in them. It was a long stressful 12 days after the birth that the couple agreed with the recommendation that they raise their baby – now a happy, confident and smart young teen – as a girl. Since then, Ellie has gone on to set up an online resource dsdfamilies.org to support parents and children affected by DSD.

In the period of uncertainty, social interactions both inside and outside the hospital can become very stressful. In most circumstances when a newborn baby needs special care, parents call on friends and family for support. With DSD, often the opposite happens and parents isolate themselves, with midwives often being the only contact they have. Ellie says that midwives can be a source of great comfort and support without being experts in DSD.

The brochure gives examples of what parents can say to friends and family, such as: 'Our baby has a hormone-related condition that affects the genital development and we cannot tell you yet

TAKE-HOME MESSAGES FOR MIDWIVES

- ✓ **DO** reassure families that they are not alone going through this experience
- ✓ **DO** engage with the family and baby, using gender-free language, such as 'little one' or 'baby'
- ✓ **DO** acknowledge that it can be difficult to manage the uncertainty
- ✓ **DO** offer them an online or hard copy of the DSD brochure
- ✗ **DON'T** offer an opinion on what the gender is likely to be
- ✗ **DON'T** avoid contact with the family

whether the baby is a boy or a girl,' or: 'It was a stressful birth, and we would be grateful for some privacy.'

Ellie says that midwives are in a good position to shield the parents within the hospital setting.

While she understands the need to avoid giving the family conflicting information before all test results are back, Ellie says that this need not mean shunning the new parents. Midwives with a confident and relaxed attitude can make a fundamental difference.

No matter what gender, a newborn with DSD has the same basic needs as every other baby. Midwives can help new parents focus on this and help them and their baby with support on feeding, nappy changing, bathing and cuddling.

Avoiding pronouns is helpful, instead use 'your baby', 'little one' or simply 'baby'. The new parents are embarking on a lifelong journey on managing DSD, their child and the outside world's lack of knowledge – Ellie says midwives can help set the tone.

'Midwives are at the starting point and if they can project acceptance, then it sets the parents up for the long term. They can make a profound difference between parents feeling like they are facing trauma and feeling empowered and having a "can-do" approach.' **M**

For more information, or online copies of the brochure, go to dsdfamilies.org or email info@dsdfamilies.org

Please email *Midwives* with your experiences of DSD at hollie@midwives.co.uk

HOW IT OCCURS

DSD occurs on a spectrum – sex development has taken a different pathway at some point for some reason. This means the appearance of the genitals can vary.

