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CURRENT AWARENESS SERVICE

Valproate: scandals, guidance and future work

The issue around prescribing sodium valproate in women and girls of childbearing age has recently seen a lot of attention from government organisations and the media. It has been the subject of new research and discussions at scientific meetings, and has resulted in many changes in guidance over the last few years. This article aims to discuss the issues surrounding valproate, update on the current guidance for prescribing this medicine and mention risks of teratogenic effects with other AEDs.

Sodium valproate was licensed in the UK in 1972 and was marketed in 1974 for general prescription. It has been used for decades to treat epilepsy and bipolar disorder. In some people, it may be the most effective anti-epileptic drug (AED) for their type of epilepsy and particularly the genetic generalised epilepsies. However, it also carries a one in 10 risk of physical birth abnormalities in babies born to women taking the AED, and a four in 10 risk of developmental and learning problems [GOV.UK, 2021]

A safety data sheet, published in the 1980-81 'Association of British Pharmaceutical Industry (ABPI) Data Sheet Compendium', referenced teratogenic effects seen in animal models with valproate medicines [Reckitt-Labaz, 1981]. It suggested that the benefits and the hazards the drug presents should be weighed up in women of childbearing age. Research literature in the 1980s was also beginning to confirm these findings, identifying cases and the characteristics of foetal valproate syndrome (FVS) [For example Tein and MacGregor, 1985; DiLiberti *et al*, 1984; Ardinger *et al*, 1988].

However, over the years, patient reports and safety reviews show that these risks were not communicated effectively to patients, leading to an estimated 20,000 people affected by exposure to sodium valproate during pregnancy [Independent Medicines and Medical Devices Safety Review, 2020].

Despite this existing evidence and concerns raised by women about the harmful effects of these medicines to their children, no specific, additional safety measures were put in place for over 40 years. Patient groups, such as In-FACT (Independent Fetal Anti Convulsant Trust), and epilepsy organisations including Epilepsy Action, have raised awareness and campaigned over the years for

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change around the use of sodium valproate. Understandably, people affected by the medicine wanted acknowledgement of their suffering and for responsibility to be duly taken. The years of silence and slow response by the government and the healthcare system have been a large contributing factor to the level of the scandal, the number of families affected and the hardships they've experienced.

From around 2015, changes to guidance and regulation began to be put in place to try to avoid further impact from taking valproate in pregnancy. In 2015, the Medicines and Healthcare products Regulatory Agency (MHRA) issued new guidance urging better communication between medical professionals and patients. In 2016, the MHRA created the 'Valproate Toolkit', designed to facilitate conversations between doctors and patients about the risks of valproate. [GOV.UK, 2016]

In July 2017, the European Medicines Agency (EMA) carried out a medicine review on valproate medicines, which included a public hearing [European Medicines Agency, 2017]. Evidence was heard from people who were directly and indirectly affected by FVS, as well as different organisations and healthcare professionals. Following this review, the EMA's Pharmacovigilance Risk Assessment Committee (PRAC) found that new measures should be put in place to avoid its use in pregnancy. In October 2017, a UK parliamentary debate was held which also highlighted the concerns around valproate use in women of childbearing potential, and suggested a public enquiry should be held.

This led to the UK government launching a safety review in 2018, of what it called three "public health scandals" – the pregnancy test drug Primodos, vaginal mesh implant and sodium valproate [Epilepsy Action, 2018]. All three of these medical products had been seen to cause life-changing problems for women and their families, prompting the review, led by Baroness Julia Cumberlege. In the same year, the MHRA strengthened prescription guidance of sodium valproate, banning its use in women and girls of childbearing potential without putting in place a pregnancy prevention plan.

The current MHRA guidelines state [GOV.UK, 2021(a)]:

- Valproate must not be used in any woman or girl able to have children unless there is a pregnancy prevention programme (PPP) in place
- The PPP includes a risk acknowledgement form which needs to be completed each time the treatment is reviewed, at least annually
- There is a ban on the use of valproate to treat epilepsy during pregnancy unless there is no other effective treatment available

Once these rules on prescribing valproate were put in place, some epilepsy specialists raised concerns about their restrictive nature and particularly in girls less than 10 years of age. This included the Royal College of Paediatrics and Child Health (RCPCH) and the British Paediatric Neurology Association (BPNA). Watkins *et al* [2019] argued that the Risk Assessment Form that prescribers are required to complete annually does not take into consideration particular circumstances that may create exemptions. This includes women with intellectual disability (ID), for whom it may be riskier to change AEDs if they are stable on valproate monotherapy or polytherapy including valproate. It also includes emergency circumstances and informed consent, where women are made fully aware of the teratogenic effects of valproate but they do not wish to participate in the Pregnancy Prevention Programme [Watkins *et al*, 2019]. The current Risk Acknowledgement Form that needs to be completed annually is available on the government website [Assets.publishing.service.gov.uk, 2021]. The BPNA wrote clear and very useful guidance about how to use and prescribe sodium valproate to girls and young women [BPNA, 2019]. This was updated in December 2020 on behalf of all the Royal Colleges that represented paediatrics and child health, physicians, obstetricians and gynaecologists, psychiatrists, nurses and general practitioners, as well as other organisations [Shakespeare and Sisodiya, 2020]. This revised guideline reflected minor changes made by the MHRA and changes to GMC decision-making guidelines, as well as including updated input from all contributing organisations.

Further changes needed

This fast-changing field has seen a big change in the prescription guidelines over the last six years. The publication of the 'First Do No Harm' report in 2020, based on the 2018 safety review, marked a milestone for campaigners. The findings showed what Baroness Cumberlege called "avoidable" suffering, "caused and compounded by failings in the health system itself".

The report made nine recommendations for the government to try to support people who have been affected and prevent further incidences with these medical products. Six months later, the government replied to the recommendations, accepting and rejecting some, while still considering others [GOV.UK, 2021(b)]

- **Recommendation 1**: The Government should immediately issue a fulsome apology on behalf of the healthcare system to the families affected by Primodos, sodium valproate and pelvic mesh [accepted]
- **Recommendation 2**: The appointment of a Patient Safety Commissioner who would be an independent public leader with a statutory responsibility. The Commissioner would champion the value of listening to patients and promoting users' perspectives in

seeking improvements to patient safety around the use of medicines and medical devices [accepted]

- **Recommendation 3**: A new independent Redress Agency for those harmed by medicines and medical devices should be created based on models operating effectively in other countries. The Redress Agency will administer decisions using a non-adversarial process with determinations based on avoidable harm looking at systemic failings, rather than blaming individuals [rejected]
- **Recommendation 4**: Separate schemes should be set up for each intervention – HPTs, valproate and pelvic mesh – to meet the cost of providing additional care and support to those who have experienced avoidable harm and are eligible to claim [under consideration]
- **Recommendation 5**: Networks of specialist centres should be set up to provide comprehensive treatment, care and advice for those affected by implanted mesh; and separately for those adversely affected by medications taken during pregnancy [work is being done on this for people affected by implanted mesh, but is still under consideration for those affected by drugs taken during pregnancy]
- **Recommendation 6**: The Medicines and Healthcare products Regulatory Agency (MHRA) needs substantial revision particularly in relation to adverse event reporting and medical device regulation. It needs to ensure that it engages more with patients and their outcomes. It needs to raise awareness of its public protection roles and to ensure that patients have an integral role in its work [accepted]
- **Recommendation 7**: A central patient-identifiable database should be created by collecting key details of the implantation of all devices at the time of the operation. This can then be linked to specifically created registers to research and audit the outcomes both in terms of the device safety and patient reported outcomes measures [accepted]
- **Recommendation 8**: Transparency of payments made to clinicians needs to improve. The register of the General Medical Council (GMC) should be expanded to include a list of financial and non-pecuniary interests for all doctors, as well as doctors' particular clinical interests and their recognised and accredited specialisms. In addition, there should be mandatory reporting for the pharmaceutical and medical device industries of payments made to teaching hospitals, research institutions and individual clinicians [under consideration]
- **Recommendation 9**: The Government should immediately set up a task force to implement this Review's recommendations. Its first task should be to set out a timeline for their implementation [rejected]

However, evidence has shown measures still need to go further. Epilepsy Action, together with Young Epilepsy and Epilepsy Society, has carried out surveys since 2016, looking at the level of awareness about the teratogenic effects among women with epilepsy. A survey carried out in 2016 found that 20% of women didn't know the risks of taking sodium valproate during pregnancy. Over a quarter (27%) of those taking sodium valproate said they had not had a discussion with their healthcare professional about it. In 2017, another survey of over 2,000 women found that 18% didn't know the risks of taking sodium valproate during pregnancy [Epilepsy Action, 2017]. More than a quarter (28%) of women who were currently taking sodium valproate said they had not been informed of the risks. This showed a lack of progress at that time, despite the MHRA's Valproate Toolkit. Another survey of 751 women was done in 2020 [Epilepsy Action, 2020], following implementation of a number of safety measures and guidance updates. This showed that 44% of respondents had not discussed the risks of taking valproate with their health professional in the last 12 months. Additionally, 41% of respondents said they had not signed an Annual Risk Acknowledgement Form, something which should happen at least annually, according to the MHRA.

More recently, in 2021, findings were published from a new Valproate Registry [NHS Digital, 2021], set up as part of recommendation 7 in the 'First Do No Harm' report. The first set of data from this registry found that 180 women were prescribed valproate in a month in which they were pregnant. This was out of 462 women who had conceived over the reporting period of April 2018 to September 2020. It is not clear whether this was done with informed consent from the women. It should also be said that the register did show some positive trends when it came to reducing the number of women and girls prescribed valproate. For example, there was a general decrease in prescribing prevalence of valproate to women and girls of childbearing potential over the report period. It also found that fewer females were being prescribed valproate for the first time.

It is clear that sodium valproate is now being prescribed more rarely, even to young girls less than 10 years of age. This is despite the fact that the drug is one of the most effective in controlling the seizures that characterise a number of the presumed genetic generalised epilepsy syndromes, including childhood and juvenile-onset epilepsy and juvenile myoclonic epilepsy [Mole *et al*, 2015].

Prescribing valproate to girls and young adults needs to be done with a lot of care in light of the guidance in place and the repercussions that have been seen. The discussion of pregnancy and pregnancy-testing in young people aged 12 years and above is clearly important and must be done with understanding and sensitivity. Clear

communication is essential to allow women, girls and their families, to make informed choices about their care. The findings from surveys and the valproate register do suggest that not enough conversations are being had between prescribers and women, girls and families about taking valproate.

The health of the woman is also at risk if these conversations are not being had. A report was published at the beginning of this year from Mothers and Babies: Reducing Risk through Audits and Confidential Enquiries across the UK (MBRRACE-UK). The report, entitled Saving Lives, Improving Mothers' Care 2020 found what it called a "concerning doubling" in maternal deaths due to sudden unexpected death in epilepsy (SUDEP). While the report found that pregnancy remains very safe in the UK, the number of women with epilepsy who died during or one year after pregnancy increased from 13 in 2013-15, to 22 in 2016-18. The report stated that "in many instances, these deaths are linked to inadequate medications management for those women either before or during pregnancy".

It is not unheard of for women to reduce or stop their medicines if they find out they're pregnant, for fear of harming their baby. This is associated with a great risk to the woman and her baby if she had seizures as a result. Conversations about valproate or other epilepsy medicines and pregnancy should include advice to women not to stop taking their medicine if they become pregnant, but to speak to their doctor. They should be offered preconception counselling to help navigate epilepsy medicines during pregnancy.

Going forward

Looking ahead, it's vitally important that mistakes from the past are not repeated. This means being very clear and transparent with patients about the possible risks with sodium valproate, as well as adhering to new guidelines around its prescription. However, it also extends to other AEDs where a teratogenic risk may be present.

An MHRA Public Assessment Report January 2021 suggested that other AEDs may also carry risks of teratogenicity if taken during pregnancy [GOV.UK, 2021(c)].

Compared to a background risk of 2-3% of the general population having a baby born with a physical birth abnormality, carbamazepine, phenobarbital, phenytoin and topiramate were found to carry a higher risk. For carbamazepine and topiramate it was 4-5%, for phenytoin around 6% and for phenobarbital 6-7%. Phenobarbital and phenytoin were also found to increase the risk of memory and learning difficulties.

Lamotrigine and levetiracetam were found to be safer in pregnancy. For a number of AEDs, more data were needed to draw proper conclusions. For gabapentin, pregabalin, clobazam and zonisamide, not enough data were present to reach a conclusion about risk of physical birth abnormalities. The effect on development in babies of gabapentin, oxcarbazepine, pregabalin, topiramate and zonisamide could also not be established properly due to a lack of data [GOV.UK, 2021(c)].

Because of these findings, the MHRA has said it will expand the Valproate Registry to look at prescription of all epilepsy medicines.

Finally, further research is still required to try and determine if there may be a potentially 'safe' dose of sodium valproate that can be prescribed to women during pregnancy. More research is also needed to understand if there are other factors that may be involved in the adverse effects of the drug on the foetal (developing) brain. This is because sodium valproate continues to show a greater efficacy than other AEDs in the treatment of generalised seizures and epilepsy syndromes [Mole *et al*, 2015; Marson *et al*, 2021].

Conclusions

It can be a challenge to prescribe AEDs in female patients, given the variation within epilepsy itself, as well as lifestyles, experiences and needs of different people. The prescribing of any AED must always take account of its proven efficacy in treating seizures and its use in specific epilepsy syndromes and its known adverse side-effect profile. These issues must be discussed with patients, and, in children, with their families. Helping patients make an informed choice and using a holistic approach are key elements in the provision of good medical care.

Kami Kountcheva
Co-Editor

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Epilepsy: giving the diagnosis

Richard Appleton, consultant and honorary professor in paediatric neurology, Alder Hey Children's Health Park, Liverpool and Suffolk

A child with a disability or a potentially life-long condition, including epilepsy, offers opportunities and challenges for everyone involved in their care, as well as for the child themselves. For a child with epilepsy, these will differ in degree depending on a few factors. These include the epilepsy syndrome or type of epilepsy (if no syndrome has been identified), the age at which it develops and how well the seizures are controlled. It is also important to note that some healthcare professionals and families would not describe epilepsy as a disability, particularly when the epilepsy is self-limiting ('benign'), or well-controlled with medication. This view may clearly be influenced if the child has additional physical or learning difficulties (or both), which may be disabling in their own right. This could be the case in four-limb cerebral palsy, autism, tuberous sclerosis complex or Dravet and Rett syndromes.

Bad news has been defined as: "Any information which adversely and seriously affects an individual's view of his or her future" [Buckman, 1992]. As with beauty, bad news is often 'in the eye of the beholder'. One cannot fully estimate its impact, and whether it really is bad news, until one has seen and heard the recipient's understanding and expectations. However, with potentially bad news, there is far more likely to be a consensus on when and how it should be given [Brouwer *et al*, 2021].

At some point, the family and the child will have to be given the diagnosis of the disability or epilepsy. Giving the diagnosis to parents and the child may be a difficult and complex task that few would admit to finding enjoyable, and some might even find uncomfortable or stressful [Fallowfield and Jenkins 2004]. When done well, it can reduce parental confusion, dissatisfaction, fear and anguish. It can also help to establish positive and important parent–professional relationships at a crucial time in what may be a long-term process [McLaughlin *et al*, 2005]. The relationship may last all the way through to transition and a gradual hand-over into adult services. Conversely, "poor communication during initial diagnosis can leave a legacy of mistrust and anger that influences future relationships between parents and the range of health and social care professionals they come in contact with" [McLaughlin *et al*, 2005]. Finally, this process should always be a dialogue, something that is fundamental to good communication, and requires listening as well as talking. It is inevitable that the child or young person and their family will have questions about the diagnosis, and its implications, management and prognosis, and these must be answered.

An editorial in the *British Medical Journal*, published over thirty years ago, entitled 'It isn't epilepsy is it, doctor?' focused on the importance of establishing a correct diagnosis of epilepsy using clinical (not EEG) criteria [Brett, 1990]. However, the anxiety – implicit, if not actually audible – in the question, 'It isn't epilepsy is it, doctor?' highlights the negative emotions and even fear inherent in the diagnosis of epilepsy. These fears reflected, in part, the belief among parents that a child could die in a febrile convulsion [Baumer *et al*, 1981]; and clearly this could equally apply to an epileptic convulsion. A secondary fearful belief was that if their child survived, they would have suffered brain damage and consequent learning difficulties. Fortunately, this fear, founded in the 19th and perpetuated in the 20th century through ignorance and stigma, has been greatly reduced for many reasons:

- An increased understanding of the nature of epilepsy in being neither a contagious nor a mental illness
- The different syndromes and types of epilepsy, their causes and prognoses
- The discovery of more effective and more patient-friendly anti-epileptic medications
- Surgery as a much more feasible, available and early treatment option
- The support of epilepsy specialist nurses
- The role and support of the voluntary sector
- Reduced societal discrimination

Despite this welcome reduction in the mystique and fear about epilepsy, it remains important that the initial disclosure of its diagnosis is clear, knowledgeable and honest, while also being realistic and empathetic.

A paper, published in *Seizure* almost 20 years ago sought opinions on how epilepsy was disclosed to families by paediatric neurologists throughout the UK. The authors had the hope, if not expectation, that this might lead to a consensus statement on 'best practice' in this area [Cunningham *et al*, 2002]. This was a questionnaire-based survey sent to 32 consultant paediatric neurologists, which represented approximately 75% of those in practice in the UK in 2000/1. Sixteen (50%) questionnaires were completed.

Seven recurring factual points emerged from the brief written accounts within the completed questionnaires.

1. It was important to say it is epilepsy
2. There was the need to explain that seizures and fits and

Richard Appleton, consultant and honorary professor in paediatric neurology, Alder Hey Children's Health Park, Liverpool and Suffolk

some convulsions are all the same (*clearly, in 2021 this requires qualification*)
3. That recurrent seizures are called epilepsy – and that this is all that epilepsy means (*in 2021 it is probably more appropriate to say that epilepsy is defined as having a risk of experiencing recurrent seizures*)
4. Seizures can happen to anyone in certain circumstances
5. Having a seizure is very common – 'to try to counteract the connotations of the term epilepsy'; 'the brain is still working all right and is not usually damaged by the seizure'
6. It may not be persistent (depending upon the type)
7. There are different types of epilepsy with different implications or prognosis

Predictably, the results were heavily influenced by the questionnaire used in the survey which was not perfect. Many respondents did not complete every section and some complained that a questionnaire approach was too 'constraining'. Four respondents stated this was the reason they would not complete the questionnaire. The most striking impression from all the replies was the general lack of consensus in both the thoughts of, and approaches to, the disclosure of the diagnosis. The respondents considered that intuition, rather than a shared knowledge of the processes involved, determined the pattern of the disclosure. Some felt that individual improvisation and intuition based on experience was the only practical approach. Others used a clear and pre-determined pattern and procedure of what should and should not be done, including the use of other resources. It must be emphasised that, at that time, many of the respondents did not have an epilepsy specialist nurse as part of their epilepsy service and this will almost certainly have influenced some responses.

Most, but not all, did not feel all the necessary information (eg. on lifestyle, education and potential career choice), could, or should, be given at the initial clinic visit when the diagnosis was first disclosed. Mortality, including sudden unexpected death in epilepsy (SUDEP), was not listed in the questionnaire as a specific topic to be discussed. This, again, was a sign of the times, when it was considered neither necessary nor comfortable to discuss SUDEP. The National Sentinel Clinical Audit on epilepsy-related death was only published slightly later, in May 2002 [Hannah *et al*, 2002].

Overall, the survey showed that respondents used one of three approaches to give information to parents and families. These were: 'proactive', 'reactive' and 'drip-feed'.

The paper concluded with the following: 'Our aim in carrying out this survey was to find consensus from which to establish agreed guidelines. Clearly the results indicate that this is still some way off''. The lack of consensus was felt to reflect three things:

- The method of information gathering (an imperfect questionnaire)
- The heterogeneity of the epilepsies (which precluded a single or 'one size fits all' approach to disclosure)
- The status of disclosure practice at that time

This latter point would also have reflected the experience and the personality of the consultant disclosing the diagnosis. Finally, it should be emphasised that the study's conclusions were based on only 16 respondents, half of those originally contacted.

There have been few other publications on how to disclose or give a diagnosis of epilepsy, to any age group. This is in contrast to a large number of papers on how to give a diagnosis of cancer or, to a far lesser extent, of a serious or progressive neurological disorder. A study published in 2000 outlined a six-stem algorithm in how to give a diagnosis of a cancer [Baile *et al*, 2000]. This was given the acronym, 'SPIKES', which represented:

- **S**etting the interview
- Assessing the patient's **p**erception
- Obtaining the patient's **i**nvitation
- Giving **k**nowledge and information to the patient
- Addressing the patient's **e**motions with empathic responses
- **S**trategy and summary

A recent paper, published in early 2020, undertook a 'scoping review' of studies on the diagnosis-disclosure to adults with motor neurone disease, multiple sclerosis and Parkinson's disease, all progressive neurological diseases [Anestis *et al*, 2020]. The authors identified 47 studies for their review. Although patients were generally satisfied with how the diagnosis was given, a considerable proportion was still dissatisfied with aspects of the consultation. These were particularly the information given, the time provided (ie. the duration of the consultation) and the doctor's approach (specifically the lack of empathy). Only six of the 47 studies addressed doctors' perspectives, which focused more on doctors' practice. The authors concluded that although basic standards of good practice were being met, a significant proportion of patients were dissatisfied with how their diagnosis was given to them. It was considered that all healthcare professionals who have to give a diagnosis of a serious and progressive neurological disease need to:

- Assess and respond to patients' information needs
- Provide time for questions
- Maintain an empathic attitude

One could easily argue that this represents common sense, good communication and, more broadly, the art of medicine.

Interestingly, there is slightly more literature on how the

families of children with a new diagnosis of epilepsy themselves then proceed to disclose and share the diagnosis with others outside the immediate family. This includes grandparents, friends, teachers and others who may at some point be involved with the children (eg. during school or out-of-school activities). A study from Ireland, published in 2017, explored the challenges parents of children with epilepsy experienced when deciding to disclose their child's epilepsy diagnosis to others [Benson *et al*, 2017]. The authors used a qualitative exploratory design and conducted interviews with 34 parents (27 mothers and seven fathers) of 29 children aged six to 16 years. The families were recruited from a neurology clinic of a specialist children's hospital and from a national epilepsy association. The results showed five themes that represented the main challenges that families identified which either led to concealment of the diagnosis or sharing only some aspects of the diagnosis:

- Seeking or trying to maintain normalcy for the child
- The invisibility of epilepsy
- Negative reactions to disclosure
- Dealing with poor public perceptions of epilepsy
- Coming to terms with the diagnosis themselves

The authors rightly concluded that this information should help healthcare professionals to recognise families' concerns. It should also provide them with the support and resources to help families, and the children themselves, to give or share the diagnosis of epilepsy. In so doing, this would help the families' own adjustment and psychosocial wellbeing. Clearly, this was only a single study with a small number of participants and from a single country. However, an earlier systematic review of 17 studies published by the same group [Benson *et al*, 2015], reported broadly similar findings. They identified the important barriers to disclosure were:

- Prior negative responses to disclosure
- Parental fear of:
  - Stigmatisation
  - Their child being treated differently
  - Imposition of unnecessary restrictions on their child

Cultural, religious and societal issues are likely to be important factors on the disclosure of epilepsy and its wider discussions, particularly in the role and effect of stigma. We must also acknowledge that stigma may be real but also perceived by the child or family. Both need to be addressed to facilitate children's and their family's understanding, acceptance of, and adjustment to the condition. In my 26 years as a consultant, I have cared for many hundreds of children with epilepsy, most with complex and refractory epilepsy. I can recall only a handful of families that specifically asked for my advice on with whom they should share the diagnosis and when and how this should be done. However, many more asked this question of the epilepsy specialist nurses at subsequent clinic visits. They all shared the concern that the children might be excluded from a range of activities or 'isolated' if the diagnosis became more widely known. Sadly, this still remains the case in some areas of the UK and also other countries. As healthcare professionals, we must be comfortable about how we give a diagnosis of epilepsy before we are then able to help families, children and young people to do the same. If we have difficulties in disclosing the diagnosis, or give it in a piece-meal, awkward or emotionally-charged way, this may adversely impact the family's desire and ability to disclose the diagnosis to others.

It is often eye-opening (even jaw-dropping) and salutary when we have the rare opportunity to 'sit on the other side of the desk', not as a doctor or nurse, but as a patient, parent or grandparent. It provides a valuable perspective, if not insight, when we are the recipients of a diagnosis that may have life-long or life-shortening consequences on ourselves or a family member. On returning to the safety and comfort of our usual side of the desk, we should not forget our own uncomfortable and perhaps distressing experiences. We should draw on what we learned from these when we disclose the diagnosis of epilepsy to the next family we see.

In the current and COVID-19-enforced climate of virtual and online consultations, there is a risk that 'telehealth' may become the default form of consultation at the expense of the face-to-face consultation. It is my opinion that all healthcare professionals should strive to ensure the disclosure of a diagnosis of epilepsy be done in person and face-to-face. I cannot conceive of any scientific or public health reason to do otherwise and particularly from mid to late-2021 onwards.

I consider there are a few fundamental yet crucial hooks on which to hang a disclosure of epilepsy. Clearly, families may react differently to the diagnosis, and this will have to be reflected in some flexibility as to how and when it is given. Although the following hooks may seem intuitive, if not obvious to many, this may not apply to everyone. This may be particularly so for those inexperienced in disclosing diagnoses and particularly when the epilepsy syndrome or its cause is likely to be severe and difficult to control.

1. <u>Allow adequate time to undertake the disclosure</u>
   It is difficult, if not impossible and perhaps even inappropriate, to prescribe a specific duration over which this should be done. This is because of the heterogeneity of the epilepsies, the underlying cause (if known) and the response of individual families to the disclosure. However, my experience would suggest a minimum of 45 and ideally 60 minutes is a reasonable time. This allows time to give the diagnosis and discuss the immediate management plan and relevant lifestyle issues. Many families are likely to require a second appointment, probably within a few weeks after the initial disclosure. This helps to 'retrace their steps' in what might have

Richard Appleton, consultant and honorary professor in paediatric neurology, Alder Hey Children's Health Park, Liverpool and Suffolk

been a challenging and confusing initial disclosure, or allows them the opportunity to ask questions that were not addressed or asked at the first appointment. Clearly, many families will probably need both.

2. Seek a family's understanding of epilepsy before this is explained in detail
   An early and brief exploration of 'where the family are' in their understanding of their child's symptoms and subsequent diagnosis is important. It enables the clinician to identify the child's and family's fears but also any myths and misunderstandings if they already believe their child has epilepsy. It also allows the family to share with the doctor what they have found from the internet; this is likely to reflect the family's postcode and background. This knowledge will enable a more appropriate and individual approach to the disclosure. It also shows the family that the doctor is aware of the importance of communication as a dynamic, two-way dialogue. This question-and-answer approach should continue throughout the consultation (and beyond), to ensure the family and child can proceed at a rate with which they are comfortable.

3. Speak honestly with the family and child but framed with optimistic realism
   This emphasises the importance of knowing about epilepsy and specifically the different seizure types and syndromes, and possible underlying causes (particularly a genetic disorder or a potentially surgically-treatable lesion). It's important to also know the available treatment options based on their efficacy and safety data, and the likely prognosis for the child. There will always be some children in whom it may not be immediately clear what epilepsy syndrome they have, or its cause. In these situations, discussing uncertainty with the family is important, because it is true. A specialist in paediatric epilepsy from a tertiary centre must be involved if there is any doubt over the initial diagnosis of epilepsy or the specific epilepsy syndrome [NICE 2012]. Providing inaccurate, skewed or inappropriately pessimistic or optimistic information represents poor clinical medicine. The situation is further compounded if the inaccurate or skewed information is given in an engaging, eloquent and charming style and in what seems to be an excellent 'bed-side manner'. Dr Richard Asher, in his insightful book, 'Talking Sense', wrote: "It is a greater medical triumph to leave the patient feeling better, but thinking little of the doctor, than to leave him worse, but deeply impressed" [Asher, 1972]. Clearly, the ultimate goal and triumph would be to have the patient feeling better and deeply impressed with the doctor. A good doctor recognises the importance of both the science (a sound knowledge and understanding of the condition) as well as the art (how to communicate this knowledge and understanding) of medicine. Focusing on the art while ignoring or misrepresenting the science risks leading the child and family into a false sense of security and trust. If this becomes a reality, it may result in a significant adverse long-term impact on their relationship with the doctor, if not the wider medical profession.

4. Be sensitive in the disclosure
   As doctors and nurses, we are encouraged not to become emotionally involved with our patients. This can be difficult to avoid when we have known the child and the family for many years, and over a period of time during which everyone has grown together. Clearly, emotional involvement is far less likely in the early stages of a potential long-term doctor-patient journey. However, this does not mean we should not show empathy and sensitivity when we talk with the child and their family. This is when, as we begin the disclosure consultation, we should ask ourselves: 'How would we like to be told our son or daughter has epilepsy and what this might mean for them, their siblings and ourselves as parents?' Finally, we need to view and treat the child as a person with epilepsy and not as a person whose seizures we must control, as this will not always be possible. We should keep in focus what the children themselves think, "I don't want them to look at me and think of my illness, I just want them to look at me and see me" [Benson *et al*, 2015). This clearly emphasises treating patients (of all ages) holistically.

5. Provide the family with literature and details of websites from where they can obtain more information
   It is very likely – probably inevitable – that families will want confirmation and further information on what they heard during the consultation, even though they may initially say they don't need it. This is irrespective of the duration of the initial consultation and the lack of any questions about the diagnosis and its implications at that time. Some will have been too shocked or not have known which questions to have asked at the disclosure visit. It is not appropriate to simply suggest the family go to the internet or consult Dr Google, because of the risk they will find inappropriate or confusing information. I also don't consider that they should rely on a copy of the clinic letter, as the family may not receive this for some weeks (possibly longer), and it is likely to contain medical jargon. All epilepsy clinics should provide families with printed information sheets on the relevant epilepsy syndrome and anti-epileptic medication(s). Information should include details of reliable and up-to-date websites from where they can obtain additional information (such as from organisations including Epilepsy Action). I believe it is important that the families leave the clinic with some of this 'real' (rather than virtual) information, even if this is limited. This should certainly be possible in the gradually easing of the COVID-19 restrictions.

Patient experiences of any consultation are clearly important and it is an area that was picked up and explored by the Picker Institute over 15 years ago. The Picker Institute is an organisation and charity that evaluates the development of valid measures to assess and gauge patient experiences. The Institute has previously worked with the Healthcare Commission in a number of projects, including an assessment of the day-case and inpatient experience of young people and their parents. Their early research identified eight domains that were considered important in patient experiences as both outpatients and inpatients [Picker Institute 2005]. These were:

1. Fast access to reliable health advice
2. Effective treatment delivered by trusted professionals
3. Involvement in decisions and respect for preferences
4. Clear information, communication and support for self-care
5. Attention to physical and environmental needs
6. Emotional support, empathy and respect
7. Involvement and support for family and carers
8. Continuity of care and smooth transitions

It is perhaps surprising that there was no specific domain on diagnosis and specifically disclosing a diagnosis of a serious and potentially life-long or life-shortening disease. It is possible this was considered too narrow a subject and therefore did not justify a specific domain. However, I would argue that the initial disclosure of a diagnosis is a very important, if not crucial, first step in the building and development of a patient (and family) experience.

Numerous attempts and using different instruments have assessed the doctor-patient consultation, the vast majority of which have been in adults [Crossley *et al*, 2005; Sitzia, 1999]. A more recent, but, again, small study in the US, which involved only 28 physicians and students, focused on how to help medical staff improve breaking bad news to children and their families [Kukora *et al*, 2020]. Twenty four of the 28 (86%) considered that the training programme was effective.

A national pilot study published in 2011 by the Royal College of Paediatrics and Child Health (RCPCH) identified a 'valid and reliable method of carer feedback' on a doctor's performance [McGraw *et al*, 2011]. I took part in this pilot study. The study used a 17-item questionnaire that was based on the Sheffield Patient Assessment Tool (SHEFFPAT). This tool was identified by the Picker Institute as meeting their standards for content, validity and reliability in the assessment of patient consultations [Chisholm and Askham 2006]. The questionnaire was slightly modified by the RCPCH and renamed the 'Paediatric Carers of Children Feedback tool (PaedCCF). Most paediatricians now have to complete this tool as part of their annual appraisal and revalidation. The questionnaire was completed after a single and random outpatient consultation by the parents with input from the child and young person wherever possible [McGraw *et al*, 2011]. The consultation could have been a new or follow-up appointment and might have included a consultation where there had been the first disclosure of a diagnosis. It could have been regarding any medical or surgical condition, not just epilepsy. None of the 17 questions in the PaedCCF specifically included one on diagnosis or its disclosure. Two of the 17 questions which were indirectly related to the disclosure of a diagnosis were:

1. How well do you understand your child's condition now you have seen the doctor?
2. How well did the doctor explain the risks of your child's condition?

Although the Picker Institute had endorsed the SHEFFPAT tool and was therefore indirectly involved in PaedCCF, as of 2020, it has not undertaken any project on diagnosis-disclosure. In my opinion this represents a missed opportunity.

Conclusion

The disclosure of a diagnosis of epilepsy is a very important first step in the management of epilepsy in any individual. In a child, this may be more complex because the disclosure involves not only the child but their family. It may also mean 'bad news', because the epilepsy or epilepsy syndrome may prove to be life-long, progressive and difficult to treat. It may also develop in a child with pre-existing physical or learning difficulties, which some families may consider 'the final straw'. Potentially life-changing epilepsy will also impact on the child's parents and siblings [Hames and Appleton, 2009] and this may have a long-term adverse impact on their own lives.

Getting the disclosure of epilepsy right will help to reduce confusion and misunderstanding early in the child's management. It is likely to facilitate future professional doctor-child and doctor-family relationships, which is important for the 'patient experience'.

Communication is crucial to disclosing a diagnosis; it must always be a two-way process to allow the family to respond and progress and at a rate with which they are comfortable. Key factors that are important in achieving a successful disclosure-consultation are:

- Providing adequate time
- Seeking the family's prior understanding of epilepsy
- Having a sound knowledge of the epilepsy syndromes and epilepsies (and if in doubt seeking the advice of a paediatric neurologist with expertise in epilepsy)
- Discussing the epilepsy honestly and with sensitivity, and
- Sharing up-to-date and reliable information

Richard Appleton, consultant and honorary professor in paediatric neurology, Alder Hey Children's Health Park, Liverpool and Suffolk

Prof Richard Appleton, consultant and honorary professor in paediatric neurology, Alder Hey Children's Health Park, Liverpool and Suffolk

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epilepsy action

epilepsyspace.org.uk

The Epilepsy Space

The mobile friendly website is a helping hand for 16-25 year olds to live their best life with epilepsy

The Epilepsy Space will help young people to:

- Manage their epilepsy
- Feel less alone
- Increase their confidence
- Get the support they need

There's lots of epilepsy facts, tips and stories from young people sharing their experience.

The content is short and interactive. It's not all reading, there's video and young people can share their own quotes, stories and videos too.

It's been created with young people and reviewed by epilepsy nurses.

Take a look at:
epilepsyspace.org.uk

Leaflets about The Epilepsy Space to give to young people can be requested by emailing:
nurseorders@epilepsy.org.uk

Epilepsy Action
Information you can trust

Find out more
epilepsy.org.uk/trust

Recently published papers

This section highlights recently published papers. Hopefully this will be very useful to all, helping to keep everyone up to date with the latest developments. It will certainly save you research and reading time, not having to search so many journals.

There are many (often over 300) epilepsy papers published every three months, so what follows has been edited. All animal papers have been excluded and as many review papers as possible have been included. We hope you find the papers of interest in your pursuit to keep abreast of the very latest knowledge.

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New Anstey House, Gate Way Drive, Yeadon,
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