

## Supplement 6 - Results of Round 2 Voting

Dear All

Thank you for voting on the guidelines in the first round. For guidelines that reached less than 80% agreement (i.e. less than 80% voted 'yes') we have, based on your feedback, adapted the guidelines so that they can be voted on again. This time there are only two option 'Yes' (Agree with guideline) or 'No' (Don't agree with the guideline).

Thanks for helping with this

Colin Reilly  
Chair ILAE paediatric Psychiatric Issues Task Force.

### NICE guidelines

<b>Local pathway for recognition, referral and diagnostic assessment of possible autism</b>	Yes	No
1. Each region, where feasible, should have one body – the local autism strategy group - responsible for the planning and implementation of assessment of autism in children with epilepsy.	90%	10%
2. The local autism strategy group should appoint a lead professional to be responsible for the local autism pathway for recognition, referral and diagnosis of children and young people with epilepsy. The aims of the group should include: <ul style="list-style-type: none"> <li>• improving early recognition of autism in children with epilepsy by raising awareness of the features suggesting possible autism through multi-agency training (see appendix 1, 2 and 3)</li> <li>• making sure the relevant professionals (healthcare, social care, education and voluntary sector) are aware of the local autism pathway and how to access diagnostic services.</li> <li>• supporting the smooth transition to adult services for young people with epilepsy going through the diagnostic pathway</li> <li>• ensuring data collection and audit of the pathway takes place.</li> </ul>	90%	10%
3. In each area where feasible a multidisciplinary group (the autism team) should be set up. The core membership ideally should include or have regular access to the following if they are not in the team: <ul style="list-style-type: none"> <li>• Pediatric neurologist or pediatrician with expertise in epilepsy</li> <li>• Speech and language therapist</li> <li>• Occupational therapist</li> <li>• Psychologist with training and experience in assessing and monitoring autistic children and young people.</li> <li>• Child and adolescent psychiatrist</li> <li>• Developmental pediatrician with expertise in autism.</li> </ul>	100%	0%
4. Families of the child with epilepsy should have a clear point of contact within the autism team during the assessment process	100%	0%
5. The autism team should either have the skills (or have access to professionals that have the skills) needed to carry out an autism diagnostic assessment, for children and young people with epilepsy but also other coexisting conditions including:	100%	0%

<p>a. severe visual and hearing impairments, motor disorders including cerebral palsy, severe learning (intellectual) disabilities, complex language disorders or complex mental health disorders.</p> <p>b. looked-after children and young people (i.e. children not in the care of the parents who are being looked after by social services)</p>		
<b>Recognising children and young people who may be autistic</b>	<b>Yes</b>	<b>No</b>
6. Consider the possibility of autism if there are concerns about development or behaviour, but be aware that there may be other explanations for individual signs and symptoms including overlap with epilepsy other neurodevelopmental/neurogenetic conditions and psychosocial issues.	100%	0%
<b>Referring children and young people to the autism team</b>	<b>Yes</b>	<b>No</b>
7. Children with epilepsy who experience a regression in language, motor or social skills should be referred for an autism assessment	100%	0%
<p>8. When referring children and young people with epilepsy to the autism team, include in the referral letter the following information:</p> <ul style="list-style-type: none"> <li>• Reported information from parents, carers and professionals about signs and/or symptoms of concern</li> <li>• Your own observations of the signs and/or symptoms</li> <li>• Information about the child's epilepsy including age of seizure onset, seizure types, treatment and previous and ongoing epilepsy related investigations</li> <li>• Antenatal and perinatal history</li> <li>• Developmental milestones</li> <li>• Factors associated with an increased prevalence of autism (see Appendix 4)</li> <li>• Relevant medical history and investigations</li> <li>• Information from previous assessments.</li> </ul>	100%	0%
9. If you do not think concerns are sufficient to prompt a referral and you feel you are qualified to make this judgement, consider a period of watchful waiting. If you remain concerned about autism, reconsider your referral decision.	100%	0%
10. If the parents or carers or if appropriate, the child or young person with epilepsy, prefer not to be referred to the autism team, consider holding a meeting of all stakeholders (e.g. family, medical professionals) to discuss the rationale for referral, and advantages/disadvantages of onward referral for an autism assessment.	100%	0%
11. If a concern has been raised but there are no features suggesting possible autism or other reasons to suspect autism, convene a meeting of all stakeholders to discuss the next steps.	100%	0%
12. Start the autism diagnostic assessment as early as possible and within 3 months of the referral to the autism team if possible.	100%	0%
13. A case coordinator/worker/clinician in the autism team should be identified for every child or young person with epilepsy who is to have an autism diagnostic assessment.	100%	0%
14. The autism case coordinator/worker/clinician should ideally:	100%	0%

<ul style="list-style-type: none"> <li>• act as a single point of contact for the parents or carers and, if appropriate, the child or young person with epilepsy being assessed, through whom they can communicate with the rest of the autism team</li> <li>• keep parents or carers and, if appropriate, the child or young person, up-to-date about the likely time and sequence of assessments</li> <li>• arrange the provision of information and support for parents, carers, children and young people as directed by the autism team</li> <li>• gather information relevant to the autism diagnostic assessment including details about the child’s epilepsy and treatment</li> </ul>		
<p>15. Consider the following differential diagnoses for autism and whether specific assessments are needed to help interpret the autism history and observations but also :</p> <ul style="list-style-type: none"> <li>• Neurodevelopmental disorders: <ul style="list-style-type: none"> <li>○ specific language delay or disorder</li> <li>○ a learning (intellectual) disability or global developmental delay</li> <li>○ developmental coordination disorder (DCD).</li> </ul> </li> <li>• Mental and behavioural disorders: <ul style="list-style-type: none"> <li>○ attention deficit hyperactivity disorder (ADHD)</li> <li>○ mood disorder</li> <li>○ anxiety disorder</li> <li>○ attachment disorders</li> <li>○ oppositional defiant disorder (ODD)</li> <li>○ conduct disorder</li> <li>○ obsessive compulsive disorder (OCD)</li> <li>○ psychosis.</li> </ul> </li> <li>• Other conditions: <ul style="list-style-type: none"> <li>○ severe hearing impairment</li> <li>○ severe visual impairment</li> <li>○ maltreatment</li> <li>○ selective mutism</li> </ul> </li> </ul>	100%	0%
<p>16. Be aware that in some children and young people with epilepsy there may be uncertainty about the diagnosis of autism, particularly in:</p> <ul style="list-style-type: none"> <li>• children with epilepsy with very frequent seizures or who have a Developmental and Epileptic Encephalopathy</li> <li>• children with epilepsy younger than 24 months</li> <li>• children or young people with epilepsy with severe/profound ID or a developmental age of less than 18 months</li> <li>• children or young people with epilepsy for whom there is a lack of available information about their early life (for example, some looked-after or adopted children)</li> <li>• older teenagers with epilepsy</li> <li>• children or young people with epilepsy who also have a complex coexisting mental health disorder (for example ADHD, conduct disorder, a possible attachment disorder), sensory impairment (for example severe hearing or visual impairment), or a motor disorder such as cerebral palsy.</li> </ul>	100%	0%

<p>17. Consider whether the child or young person with epilepsy may have any of the following as a coexisting condition, and if suspected carry out appropriate assessments and referrals:</p> <ul style="list-style-type: none"> <li>• Mental and behaviour problems and disorders: ADHD Anxiety disorders and phobias Mood disorders Oppositional defiant behaviour Tics or Tourette syndrome, OCD, Self-injurious behaviour.</li> <li>• Neurodevelopmental problems and disorders: Global delay or a learning (intellectual) disability, Motor coordination problems or DCD Academic learning problems, for example in literacy or numeracy, Speech and language disorder.</li> <li>• Medical or genetic problems and disorders: Chromosome disorders Genetic abnormalities, including fragile X, Tuberous sclerosis Muscular dystrophy, Neurofibromatosis.</li> <li>• Functional problems and disorders: Feeding problems, including restricted diets Urinary incontinence or enuresis Constipation, altered bowel habit, faecal incontinence or encopresis Sleep disturbances Vision or hearing impairment.</li> </ul>		
<b>Medical Investigations</b>	<b>Yes</b>	<b>No</b>
<p>18. Do not routinely perform any medical investigations as part of an autism diagnostic assessment, but consider the following in individual circumstances and based on physical examination, clinical judgment and the child or young person's profile:</p> <ul style="list-style-type: none"> <li>• genetic tests, as recommended by your regional genetics centre, if there are specific dysmorphic features, congenital anomalies and/or evidence of an intellectual disability.</li> </ul>	90%	10%
<b>Communicating the results from the autism diagnostic assessment</b>	<b>Yes</b>	<b>No</b>
<p>19. Share information, including the written report of the diagnostic assessment, with the child's primary health care professional/physician</p>	100%	0%
<p>20. For children and young people with epilepsy who have a diagnosis of autism, it may be useful to discuss with parents or carers the risk of autism occurring in siblings and future children, but this discussion will also depend on the nature of the child's epilepsy and medical history.</p>	100%	0%

The features suggesting possible autism are a combination of delay in expected features of development and the presence of unusual features, and are intended to alert professionals to the possibility of autism in a child or young person about whom concerns have been raised. They are not intended to be used alone, but to help professionals recognise a pattern of impairments in reciprocal social and communication skills, together with unusual restricted and repetitive behaviours.

## Appendix 1 Features suggesting possible autism in preschool children (or equivalent mental age)

### Social interaction and reciprocal communication behaviours

#### Spoken language

- Language delay (in babble or words, for example less than ten words by the age of 2 years).
- Regression in or loss of use of speech.
- Spoken language (if present) may include unusual:
  - non-speech like vocalisations
  - odd or flat intonation
  - frequent repetition of set words and phrases ('echolalia')
  - reference to self by name or 'you' or 'she/he' beyond 3 years.
- Reduced and/or infrequent use of language for communication, for example use of single words although able to speak in sentences.

### **Responding to others**

- Absent or delayed response to name being called, despite normal hearing.
- Reduced or absent responsive social smiling.
- Reduced or absent responsiveness to other people's facial expressions or feelings.
- Unusually negative response to the requests of others (demand avoidant behaviour).
- Rejection of cuddles initiated by parent or carer, although may initiate cuddles themselves.

### **Interacting with others**

- Reduced or absent awareness of personal space, or unusually intolerant of people entering their personal space.
- Reduced or absent social interest in others, including children of his/her own age – may reject others; if interested in others, may approach others inappropriately, seeming to be aggressive or disruptive.
- Reduced or absent imitation of others' actions.
- Reduced or absent initiation of social play with others, plays alone.
- Reduced or absent enjoyment of situations that most children like, for example, birthday parties.
- Reduced or absent sharing of enjoyment.

### **Eye contact, pointing and other gestures**

- Reduced or absent use of gestures and facial expressions to communicate (although may place adult's hand on objects).
- Reduced and poorly integrated gestures, facial expressions, body orientation, eye contact (looking at people's eyes when speaking) and speech used in social communication.
- Reduced or absent social use of eye contact, assuming adequate vision.
- Reduced or absent joint attention shown by lack of:
  - gaze switching
  - following a point (looking where the other person points to – may look at hand)
  - using pointing at or showing objects to share interest.

### **Ideas and imagination**

- Reduced or absent imagination and variety of pretend play.

### **Unusual or restricted interests and/or rigid and repetitive behaviours**

- Repetitive 'stereotypical' movements such as hand flapping, body rocking while standing, spinning, finger flicking.
- Repetitive or stereotyped play, for example opening and closing doors.
- Over-focused or unusual interests.
- Excessive insistence on following own agenda.

- Extremes of emotional reactivity to change or new situations, insistence on things being 'the same'.
- Over or under reaction to sensory stimuli, for example textures, sounds, smells.
- Excessive reaction to taste, smell, texture or appearance of food or extreme food fads.

## **Appendix 2 Features suggesting possible autism in primary school children (aged 5 to 11 years or equivalent mental age)**

### **Social interaction and reciprocal communication behaviours**

#### **Spoken language**

- Spoken language may be unusual in several ways:
  - very limited use
  - monotonous tone
  - repetitive speech, frequent use of stereotyped (learnt) phrases, content dominated by excessive information on topics of own interest
  - talking 'at' others rather than sharing a two-way conversation
  - responses to others can seem rude or inappropriate.

#### **Responding to others**

- Reduced or absent response to other people's facial expression or feelings.
- Reduced or delayed response to name being called, despite normal hearing.
- Subtle difficulties in understanding other's intentions; may take things literally and misunderstand sarcasm or metaphor.
- Unusually negative response to the requests of others (demand avoidant behaviour).

#### **Interacting with others**

- Reduced or absent awareness of personal space, or unusually intolerant of people entering their personal space.
- Reduced or absent social interest in people, including children of his/her own age – may reject others; if interested in others, may approach others inappropriately, seeming to be aggressive or disruptive.
- Reduced or absent greeting and farewell behaviours.
- Reduced or absent awareness of socially expected behaviour.
- Reduced or absent ability to share in the social play or ideas of others, plays alone.
- Unable to adapt style of communication to social situations, for example may be overly formal or inappropriately familiar.
- Reduced or absent enjoyment of situations that most children like.

#### **Eye contact, pointing and other gestures**

- Reduced and poorly integrated gestures, facial expressions and body orientation, eye contact (looking at people's eyes when speaking) and speech used in social communication.
- Reduced or absent social use of eye contact, assuming adequate vision.
- Reduced or absent joint attention shown by lack of:
  - gaze switching
  - following a point (looking where the other person points to – may look at hand)
  - using pointing at or showing objects to share interest.

#### **Ideas and imagination**

- Reduced or absent flexible imaginative play or creativity, although scenes seen on visual media (for example, television) may be re-enacted.
- Makes comments without awareness of social niceties or hierarchies.

### **Unusual or restricted interests and/or rigid and repetitive behaviours**

- Repetitive 'stereotypical' movements such as hand flapping, body rocking while standing, spinning, finger flicking.
- Play repetitive and oriented towards objects rather than people.
- Over-focused or unusual interests.
- Rigid expectation that other children should adhere to rules of play.
- Excessive insistence on following own agenda.
- Extremes of emotional reactivity that are excessive for the circumstances.
- Strong preferences for familiar routines and things being 'just right'.
- Dislike of change, which often leads to anxiety or other forms of distress (including aggression).
- Over or under reaction to sensory stimuli, for example textures, sounds, smells.
- Excessive reaction to taste, smell, texture or appearance of food or extreme food fads.

### **Other factors that may support a concern about autism**

- Unusual profile of skills or deficits (for example, social or motor coordination skills poorly developed, while particular areas of knowledge, reading or vocabulary skills are advanced for chronological or mental age).
- Social and emotional development more immature than other areas of development, excessive trusting (naivety), lack of common sense, less independent than peers.

## **Appendix 3 Features suggesting possible autism in secondary school children (older than 11 years or equivalent mental age)**

### **Social interaction and reciprocal communication behaviours**

#### **Spoken language**

- Spoken language may be unusual in several ways:
  - very limited use
  - monotonous tone
  - repetitive speech, frequent use of stereotyped (learnt) phrases, content dominated by excessive information on topics of own interest
  - talking 'at' others rather than sharing a two-way conversation
  - responses to others can seem rude or inappropriate.

#### **Interacting with others**

- Reduced or absent awareness of personal space, or unusually intolerant of people entering their personal space.
- Long-standing difficulties in reciprocal social communication and interaction: few close friends or reciprocal relationships.
- Reduced or absent understanding of friendship; often an unsuccessful desire to have friends (although may find it easier with adults or younger children).
- Social isolation and apparent preference for aloneness.
- Reduced or absent greeting and farewell behaviours.
- Lack of awareness and understanding of socially expected behaviour.
- Problems losing at games, turn-taking and understanding 'changing the rules'.

- May appear unaware or uninterested in what other young people his or her age are interested in.
- Unable to adapt style of communication to social situations, for example, may be overly formal or inappropriately familiar.
- Subtle difficulties in understanding other's intentions; may take things literally and misunderstand sarcasm or metaphor.
- Makes comments without awareness of social niceties or hierarchies.
- Unusually negative response to the requests of others (demand avoidant behaviour).

### **Eye contact, pointing and other gestures**

- Poorly integrated gestures, facial expressions, body orientation, eye contact (looking at people's eyes when speaking) assuming adequate vision, and spoken language used in social communication.

### **Ideas and imagination**

- History of a lack of flexible social imaginative play and creativity, although scenes seen on visual media (for example, television) may be re-enacted.

### **Unusual or restricted interests and/or rigid and repetitive behaviours**

- Repetitive 'stereotypical' movements such as hand flapping, body rocking while standing, spinning, finger flicking.
- Preference for highly specific interests or hobbies.
- A strong adherence to rules or fairness that leads to argument.
- Highly repetitive behaviours or rituals that negatively affect the young person's daily activities.
- Excessive emotional distress at what seems trivial to others, for example change in routine.
- Dislike of change, which often leads to anxiety or other forms of distress including aggression.
- Over or under reaction to sensory stimuli, for example textures, sounds, smells.
- Excessive reaction to taste, smell, texture or appearance of food and/or extreme food fads.

### **Other factors that may support a concern about autism**

- Unusual profile of skills and deficits (for example, social or motor coordination skills poorly developed, while particular areas of knowledge, reading or vocabulary skills are advanced for chronological or mental age).
- Social and emotional development more immature than other areas of development, excessive trusting (naivety), lack of common sense, less independent than peers.

### **Appendix 4 Factors associate with an increased prevalence of autism**

- A sibling with autism.
- Birth defects associated with central nervous system malformation and/or dysfunction, including cerebral palsy.
- Gestational age less than 35 weeks.
- Parental schizophrenia-like psychosis or affective disorder.
- Maternal use of sodium valproate in pregnancy.
- A learning (intellectual) disability.
- Attention deficit hyperactivity disorder.
- Neonatal encephalopathy or epileptic encephalopathy, including infantile spasms.
- Chromosomal disorders such as Down's syndrome.

- Genetic disorders such as fragile X.
- Muscular dystrophy.
- Neurofibromatosis.
- Tuberous sclerosis.