

# Dopey's seizure

BERNARD DAN & FLORENCE CHRISTIAENS

*Neurology Department, University Children's Hospital Queen Fabiola, Brussels, Belgium*

Correspondence to: Dr B. Dan, Neurology Department, University Children's Hospital Queen Fabiola, 15 Avenue JJ Crocq, 1020 Brussels, Belgium

Angelman syndrome is a neurogenetic condition namely characterized by developmental delay, virtual absence of expressive verbal language, peculiar organization of movement, seizures and happy demeanour. This syndrome has been recognized since 1965, but it seems that Walt Disney presented an original depiction of it in his first full-length animated film, including myoclonic jerks and an apparently generalized tonic-clonic seizure.

*Key words:* Angelman syndrome; medical illustration; development; epilepsy; speech; cinema.

## INTRODUCTION

When describing current medical conditions, there has been a tendency for reviving original depictions from previous non-medical sources, such as the Bible, ancient Greek literature, medieval or Renaissance paintings. These retrospective diagnoses based on limited elements can argue in favour of the usually obvious notion that recently recognized disorders are not actually new. However, what is interesting in such descriptions, if anything, is that they were set in a culturally distinct context, therefore placing interestingly similar or dissimilar emphasis on different aspects. For example, the passage of the Bible referenced for giving an original account of the foetal alcohol syndrome warned against the dangers of paternal rather than maternal consumption of alcohol<sup>1</sup>. Such sources may also give inspiration for coining conditions' names, as for Job syndrome, Breughel syndrome, or the more recent Alice in Wonderland syndrome. In a recent issue of *Seizure*, the trails of epilepsy and syphilis were followed through Shakespeare's work and placed in a historical perspective<sup>2</sup>. In the case of Angelman syndrome, a neurogenetic condition characterized by developmental delay, absence of speech, motor impairment, happy demeanour and seizures<sup>3,4</sup>, Doctor Harry Angelman himself explained the importance of his encounter with Giovanni Francesco Caroto's painting 'Fanciullo con pupazzo':

The boy's laughing face and the fact that my patients exhibited jerky movements gave me the idea of writing an article

about the three children with a title of Puppet Children.

But neither the boy nor the puppet seemed to have Angelman syndrome. Considering the peculiar motor impairment<sup>5</sup>, it seems logical to search for original non-medical depictions of Angelman syndrome in motion pictures. Indeed, the puppet-like features<sup>3</sup> are likely to be presented in animated movies. Walt Disney's Pinocchio, for instance, was a famous happy puppet. However, he did not have any features reminding of the happy puppet syndrome. But before *Pinocchio*, in his very first and widely acclaimed full-length animated film *Snow White and the Seven Dwarfs*, Walt Disney featured a unique little fellow introduced as follows by a character appropriately named Happy:

And this is Dopey, Ma'am. He don't talk.

## DOPEY'S ANGELMAN SYNDROME

Like most patients with Angelman syndrome, Dopey has no speech but shows eagerness to communicate. In contrast to the absence of words, his understanding of speech is rather good, and he manages to express himself by efficient mimic and gesture. He has a wide smiling mouth and a marked tendency to protrude his tongue. His nose and chin are prominent. He has intermittent strabismus. He has a happy disposition, enjoys playing jokes and tricks, and he can be overwhelmingly affectionate. He tends to flap his ears when he is content, as an unusual variant to the hand flapping

classically seen in Angelman syndrome. He has a peculiar wide-based, hopping gait, and he tends to keep his elbows flexed when he walks. He has occasional myoclonic jerks and tremor facilitated by emotion. He had an apparently generalized clonic seizure while he was asleep. Although Dopey is presumably unrelated to the other dwarfs, it may be noteworthy that he is the only one with blue eyes and that his head is smaller. And when it comes to washing hands, he finds himself completely immersed in the basin, a scene which is familiar to many parents of a child who has Angelman syndrome.

## DISCUSSION

The idea that patients presenting with the constellation of signs reported by Doctor Harry Angelman in three children in 1965 had a definite condition which had not been previously described as such<sup>3</sup>, has been strongly confirmed by a large number of subsequent reports and the discovery of underlying genetic processes. Considerable insight has been offered by genetic studies of the syndrome, and a responsible gene has recently been identified on the long arm of chromosome 15<sup>6</sup>. However, current consensual diagnostic criteria still rely essentially on clinical elements<sup>7</sup>. The prevalence of Angelman syndrome is unknown, but it has been estimated to be 1 in 12 000 children<sup>8</sup>. Most cases probably go undiagnosed.

Reviewing Walt Disney's *Snow White and the Seven Dwarfs* provides an occasion for attempting a retrospective diagnosis of Angelman syndrome in Dopey, one of the film's most attractive characters. Some important anamnestic data lack, like developmental and past medical history. However, the features presented in the film are strikingly consistent with Angelman syndrome, as detailed in Table 1.

The character was not depicted in the original tale by the Grimm brothers. Despite extensive literature, documentaries and interviews on the making of *Snow White and the Seven Dwarfs*, it is difficult to get accurate information about Fred Moore's inspiration when he designed Dopey for Disney's animated film. However, we know that in his very first outlines of the film, Walt Disney had conceived seven dwarfs with distinct personalities, including one named Seventh, who was 'deaf but always listening intently, happy, spry, with quick movements'<sup>9</sup>. Early in 1936, Seventh's name changed to Dopey, as Disney said: 'To me, it's the best word I can think of for somebody who is a little off-beat'<sup>10</sup>. Later that year, the dwarfs were given voices, but Dopey was to remain forever voiceless. His personality was to be transmitted by pantomime. More anecdotal sources report that animation suggestions for keeping Dopey

from seeming too dumb included some dog-like behaviours, such as sniffing a trail. None of the available anecdotes mention inspiration from a disabled person. However, the presented animation largely fits the clinical picture of Angelman syndrome, antedating Doctor Angelman's report of three patients<sup>3</sup> by 28 years. Harry Angelman proposed to call these children 'puppet children', a term that appeared derisive or even derogatory. The name 'Dopey' is certainly even more pejorative and ill-adapted to patients with Angelman syndrome. However, the character is highly appealing, and it might serve the purpose of spreading the knowledge of Angelman syndrome among physicians and caregivers.

## ACKNOWLEDGEMENTS

We are grateful to the Walt Disney Company for their authorization to use the description of their character for medical illustration.

## NOTE

An early draft of this paper was presented as a poster at the First National Symposium on Angelman Syndrome, Brussels, November 1997.

## REFERENCES

1. Abel, E. L. *Fetal Alcohol Syndrome*. Oradell, Medical Economics Books, 1990.
2. Betts, T. and Betts, H. A note on a phrase in Shakespeare's play *King Lear*: 'A plague upon your epileptic visage' *Seizure* 1998; **7**: 407-409.
3. Angelman, H. 'Puppet' Children: A report on three cases. *Developmental Medicine and Child Neurology* 1965; **7**: 681-688.
4. Dan, B. *Le syndrome d'Angelman*. Brussels, Fondation Angelman Belgique, 1997.
5. Dan, B., Bouillot, E., Bengoetxea, A., Christiaens, F., Boyd, S. G. and Cheron, G. Motor strategies in Angelman syndrome, spastic diplegia and normal children. *European Journal of Paediatric Neurology* 1997; **1**: A3-A8.
6. Kishino, T., Lalonde, M. and Wagstaff, J. UBE3A/E6-AP mutations cause Angelman syndrome. *Nature Genetics* 1997; **15**: 70-73.
7. Williams, C. A., Angelman, H., Clayton-Smith, J. *et al.* Angelman syndrome: consensus for diagnostic criteria. Angelman Syndrome Foundation. *American Journal of Medical Genetics* 1995; **56**: 237-8.
8. Steffenburg, S., Gillberg, C. L., Steffenburg, U. and Kyllerman, M. Autism in Angelman syndrome: a population-based study. *Pediatric Neurology* 1996; **14**: 131-136.
9. Krause, M. and Witkowski L. *Walt Disney's Snow White and the Seven Dwarfs: an Art in Its making*. New York, Hyperion, 1994.
10. Hollis, R. and Sibley, B. *Walt Disney's Snow White and the Seven Dwarfs and the Making of the Classic Film*. New York, Hyperion, 1994.

Table 1: Dopey's features with respect to the diagnostic criteria for Angelman Syndrome (from 7).

| <i>Diagnostic criteria</i>   | <i>Dopey</i> |
|--|--------------|
| <b>Consistent (100%)</b>   |              |
| ☺ Developmental delay, functionally severe   | yes          |
| ☺ Speech impairment, none or minimal use of words, receptive and non-verbal communication skills higher than verbal ones   | yes          |
| ☺ Movement or balance disorder, usually ataxia of gait and/or tremulous movement of the limbs  | yes          |
| ☺ Behavioural uniqueness: any combination of frequent laughing/smiling; apparent happy demeanour; easily excitable personality, often with flapping hand movements; hypermotoric behaviour; short attention span | yes          |
| <b>Frequent (more than 80%)</b>  |              |
| ☺ Delayed, disproportionate growth in head circumference   | ?            |
| ☺ Seizures, onset usually before 3 years of age  | yes          |
| ☺ Abnormal EEG, characteristic pattern with large amplitude slow spike waves   | ?            |
| <b>Associated (20–80%)</b>   |              |
| ☺ Strabismus   | yes          |
| ☺ Tongue thrusting, sucking or swallowing disorders  | yes          |
| ☺ Feeding problems during infancy  | ?            |
| ☹ Prominent mandible   | no           |
| ☺ Wide mouth, wide spaced teeth  | yes          |
| ☺ Frequent drooling, protruding tongue   | yes          |
| ☺ Excessive chewing; mouthing behaviours   | yes          |
| ☺ Hypopigmented skin and eyes  | yes?         |
| ☺ Hyperactive tendon reflexes  | ?            |
| ☺ Uplifted, flexed arms during walking   | yes          |
| ☺ Increased sensitivity to heat  | ?            |
| ☺ Sleep disturbance  | ?            |
| ☹ Attraction to/fascination with water   | no?          |
| ☹ Flat back of head  | no           |

---

*Comments*

---

★ scarce developmental history points to lack of acquisition rather than loss of skills (e.g. 'He never tried'); socially and functionally impaired (e.g. not or marginally involved in work activities)

★ uses no words; shows some understanding of speech (e.g. when he is instructed to check the bedroom); can efficiently use own sign language (e.g. describing the 'monster')

★ coordination difficulties (e.g. the soap episode), synkinesia (e.g. turning the key of the mine door), myoclonic jerks, tremor facilitated by emotion (e.g. checking the bedroom) and a wide-based, hopping gait

★ frequent smiling; happy demeanour; ear flapping when content; easily distractable (e.g. by a fly)

★ small head best seen when the dwarfs cry over Snow White (hoods off)

★ apparently generalized clonic seizure in his sleep

★ intermittent, convergent

★ tongue thrusting

★

★

★ wide smiling mouth, only one apparent tooth

★ drooling, tongue frequently protruded

★ e.g. accidentally swallows soap (and a spoon in unreleased footages)

★ he is the only dwarf with blue eyes

★

★

★

★ has a seizure in his sleep

★ finds himself immersed in the washing basin

★

---