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ON MY MIND

Angelman Syndrome in the *Portrait of a Child With a Drawing* by Giovanni F. Caroto

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The sudden theft of invaluable paintings from the Museo Civico di Castelvecchio (Verona, northern Italy) made the headlines in Italy and around the world, causing outrage in the public. On November 19, 2015, armed men stole 17 paintings by Tintoretto, Peter Paul Rubens, Jacopo Bellini, Andrea Mantegna, and many more, whose value, according to Italian newspaper *La Repubblica*, have been estimated between €10 and €15 million (US\$11 and \$17 million).¹ Among the stolen ones, *Portrait of a Child With a Drawing* (featured in slide 7 in the slideshow in *La Repubblica*)¹ by Giovanni Francesco Caroto (ca. 1480-1555) stands out. This 1523 artwork is well known because of its artistic value and its paramount medical relevance; it had reminded the British pediatrician Harry Angelman (1915-1996) of some of his patients, who shared a certain similarity with the portrayed boy. This influenced Angelman to describe in 1965 the congenital condition, which would subsequently be named after him. The now-famous nickname *puppet children* given by him to individuals with the illness might well have originated from an erroneous translation of the word *pupazzetto*, whose primary meaning is “sketch” or “caricature,” rather than “puppet,” which is precisely the drawing the boy is holding in his hand.²

Angelman syndrome is a neurodevelopmental disorder caused by several genetic abnormalities involving chromosome 15—whose 15q11-13 region is imprinted—in 70% of cases with a de novo deletion of the maternal chromosome. Morphologically, traits such as blue eyes, fair/blond hair, elongated face, prominent jaw, large mouth, protruding tongue, flat occiput, deep-set eyes, and strabismus may be present. However, there is a great degree of variability. Characteristic of this syndrome are gait ataxia and sudden easily provoked bursts of laughter, which bring to mind the word *puppet*.³ Neuropsychologically, motor development is delayed, mental retardation is serious (IQ<25), and verbal communication is virtually nonexistent. Attentive processes lack consistency, and social development is compromised.⁴

Portrait of a Child With a Drawing was painted when Caroto was probably still active in Casale Monferrato, although his main presence there had been earlier under the protection of Marquess William IX (1486-1518)⁵; thus, it might be that the portrayed child may have been close to court life. Nonetheless, his identity is unknown and it has even been speculated that he might have been Caroto's son, as the surname is linked to the word *carrot*, whose color (ie, ginger) is the shade of the child's hair.⁶

The canvas exemplifies a new, more natural style of how children were portrayed in Renaissance art: a child acting as children would do, unlike previous depictions showing them as miniaturized adults. The sketch he holds in his hand is regarded by experts as the child's work, maybe a self-portrait. If so, because of the aforementioned neurological impairment, it would be unrealistic for the child to have had Angelman syndrome, as he would not have acquired the eye-hand coordination to draw such a shape. On the contrary, if the child did not make the drawing himself and was simply pointing to a sketch made by another, or if primarily the facial evidence is favored, the similarity between modern cases and the painting continues to appear straightforward.

Although the physical traits (a somewhat elongated face with prominence of the lower jaw, a stereotypical laughing expression, and a large mouth) are suggestive of Angelman syndrome, this contradiction limits the possibility to ascertain whether this condition really existed in the past and a cautious approach (including the possibility of seeing such patients in a figure with only vaguely similar traits) should be chosen.

In conclusion, despite certain inconsistencies and alternative interpretations of clinicopictorial correlates, seeing this painting stimulated Angelman to finalize his studies, making them available for the medical literature. This demonstrates how art and medicine are deeply interwoven.

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