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DWARFISM IN ART

KARŁOWATOŚĆ W SZTUCE

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Abstract
Throughout the history of mankind the birth of a child with congenital malformation raised anxiety and torment, along with attempts to explain its origins. It is possible to find relics of such events in prehistoric rock drawings and primitive sculptures, in numerous art pieces produced through the centuries up to modern sculptures, paintings and drawings. The aim of the present article is to show how dwarfs were portrayed in a variety of art forms at different moments in the history of our world.

Key words: achondroplasia, dwarf, sculptures, paintings, history of teratology

Streszczenie
Na przestrzeni dziejów ludzkości, narodzenie się dziecka z wrodzoną wadą rozwojową wywoływało lęk, niepokój i próby tłumaczenia nieszczęścia przez jego otoczenie. Ślady tych przeżyć znajdujemy zarówno na liczących kilka tysięcy lat rysunkach skalnych i prymitywnych rzeźbach, jak i z czasów nowożytnych, na freskach, rzeźbach, rysunkach i obrazach. Celem pracy jest przedstawienie karłowatości człowieka, której przykłady znajdujemy w dziełach sztuki pochodzących z różnych okresów rozwoju naszej cywilizacji.

Słowa kluczowe: achondroplazja, karłowatość, rzeźby, obrazy, historia teratologii

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Dwarfism is a rare medical condition that has always fascinated people and is presented in many works of art. Most of the depicted dwarfs were affected with an autosomal dominant genetic disease called achondroplasia. The cardinal features of the disorder, portrayed correctly in numerous pieces of art, include the shortening of the proximal limbs, a large head with a prominent forehead frontal bossing and a flattened nasal bridge. The worldwide incidence of achondroplasia is about 1 in 20,000 and gene penetration is 100%. It is one of the oldest phenotypically evident genetic disorders depicted in art. One of the earliest descriptions of dwarfs was made by the ancient Egyptians. The best-known is the painted statue of dwarf Seneb depicted together with his healthy wife and two children which was found in the tomb in Giza (V Dynasty, 2563-2423 B.C.) (fig. 1). He was a short-limbed dwarf with normal-shaped head, therefore some authors claim that he might rather present the hypochondroplasia and not the achondroplasia phenotype [1]. According to the tomb inscriptions, Seneb was the treasurer in the palace, where he was involved in providing clothing, supervising weaving, organizing water transport and other duties.

Dwarfs were also present at European royal courts during the Renaissance. In 1994 AEH and M. Emery provided a list of works of art where dwarfism is depicted [2]. There are records of dwarfs being present in the courts of Elizabeth I of England (1533-1603), Peter the Great of Russia (1672-1725) and the Polish King Stanisław Leszczyński (1677-1766) who had a dwarf called Nicolaus Frey ("Bebe"). Dwarfs were also present at nobleman's manor houses. For example Andrea Mantegna made a fresco of the Duke of Mantua (1474) and his court (fig. 2)[3]. The depicted individuals look around in various

Fig. 1. Egyptian Seneb with his wife and two healthy children (V Dynasty: 2563-2423) B.C.). The Egyptian Museum, Cairo, Egypt. Diagnosis – achondroplasia or hypochondroplasia [1, 3].

Fig. 2. Fresco *Duchess of Mantua and her dwarf* (1474). Artist: Andrea Mantegna. Castello di S. Giordio, Mantua, Italy. Diagnosis: achondroplasia [3].

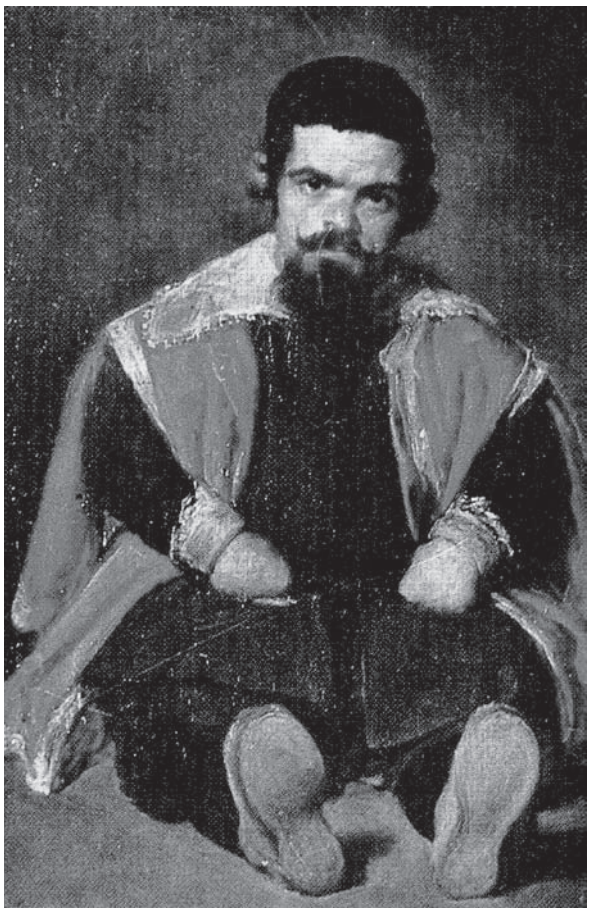

Fig. 3. Painting Sebastian de Morra. Artists: Diego Velázquez (1628). Prado Museum. Madrid. Spain [1, 3].

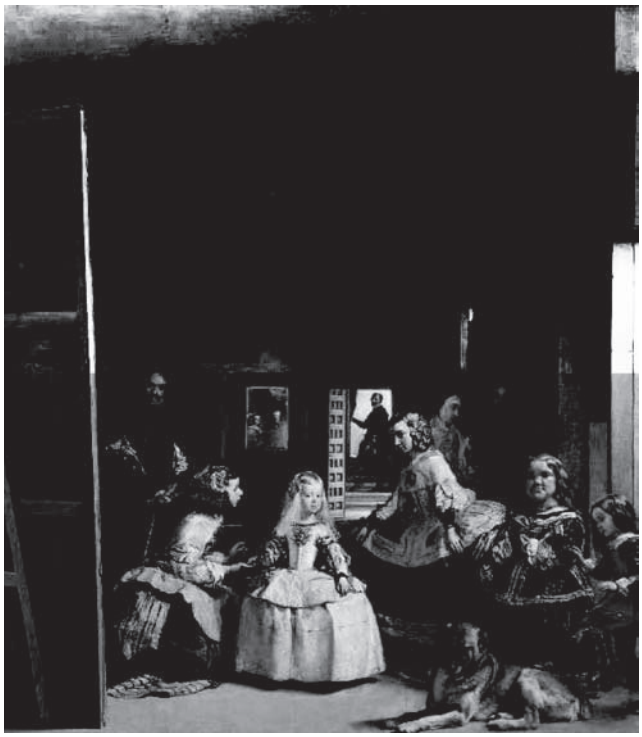

Fig. 4. Painting *Las Meninas.* Artists: Diego Velázquez (1656). Prado Museum. Madrid. Spain. In the right lower corner: Barbara Asquin ("MariaBàrbola") diagnosis: achondroplasia and Nicolaus Pertusato – STH deficiency [3, 4].

Fig. 5. Portrait of Count Jozef Boruwlaski (1739-1837). Artist unkown (1759). National Museum in Cracow, Poland. Diagnosis: dwarfism of uncertain etiology.

Fig. 6. Graphic art *Polish Dwarf*. Artist unknown – London. Count Boruwlaski with his wife and healthy daughter.

Fig. 7. Henri de Toulouse-Lautrec. Self-caricature [6].

directions except for the dwarf who glances straight at the audience. This woman, presenting typical features of achondroplasia, was probably the main subject of the painting. The Spanish painter Diego Rodriguez de Silva y Velásquez (1599-1660) painted a number of dwarfs living at the king's court. One of the portraits shows Sebastian de Morra with characteristic features of achondroplasia: a large head and shortened extremities (fig. 3). A very famous painting *Las Meninas* (The Maids of Honor) by Velásquez shows the family of Philip IV and the painter himself (fig. 4). On the right side of the painting two dwarves in the court entourage are depicted. The first is Maria Barbola who had achondroplasia: a typical large head, bulging forehead and short limbs. On her right hand stands another dwarf, Nicolaus Pertusato, presenting features characteristic for hypopituitarism. Many artists interpreted this painting, including Pablo Picasso, Francisco José de Goya and Salvatore Dali [4].

The most famous portrait of a Polish dwarf is the portrayal of count Joseph Boruwlaski (1739-1837). The author of the painting is unknown. It is exhibited in the National Museum in Cracow (fig. 5). At birth count Boruwalski was 22 cm and his adult height was finally 71 cm. There is also another drawing (*Polish Dwarf* – London 1820; fig. 6) showing the count with his wife and one of two daughters. He was well-known in the royal courts of Europe – he visited Maria Teresa and her daughter, the Queen of France, was a friend of Polish King Stanislaus August, he also knew King George III and he was a friend of George IV who sponsored his memoires (1788). He was a composer, pianist and poet

and earned a living performing his music or reading his poetry. He died at the age of 98 in Durham (England) and he wrote his epitaph:

Poland was my cradle
England is my nest
Durham is my quiet place
Where my bones shall rest [5]

Several artists suffered from genetic diseases. One of them, Henri de Toulouse-Lautrec (1864-1901), had dwarfism caused by the genetic disease pycnodysostosis (X-linked recessive disease) [6]. He created a satirical self-portrait in which the shortening of the legs is present in addition to the reddening of lips and nose (alcoholism) (fig. 7). A contemporary description of his appearance and behavior was preserved: "he had a normal torso but his rickety legs were comically shortened and his massive arms ended with huge hands with thick, shapeless twisted (…) his fragile bones broke with no reason. He limped, had huge nostrils, too large lips and his thickened tongue made it difficult for him to speak…." [6]. It is necessary to remember but it is not only the history of dwarfisms in art, but also the history of the suffering of these people [7].

Dwarfism constitutes a challenge for geneticists. Over the last decade the molecular basis of achondroplasia and hypochondroplasia were elucidated. Both diseases are caused by a defect of the receptor for fibroblast growth factor 3 (FGFR 3 gene). Two mutations, c.1138G>A (98% cases) and c.1138G>C (1-2% cases) account for more than 99% of the cases of achondroplasia. Guanine at position 1138 in the FGFR3 gene is nucleotide most predisposed for mutation identified in a human gene. Both mutations cause the same amino acid change (p.G380R) within the transmembrane domain of the receptor. It is worth nentioning that more than 80% of patients have a *de novo* mutation, while the remaining 20% are inherited from an affected parent. A *de novo* mutation occurs exclusively in the paternal germ cells and increases in frequency with the advanced age of the father. In hypochondroplasia the same gene is affected but in a different place (p.N540K). There is no causal therapy available for either disease. Growth hormone supplementation and surgical lengthening of lower legs have been proposed as treatment options of short stature. Both therapies are controversial but longitudinal studies are ongoing. The main challenge is to prevent complications of the disease such as hydrocephalus, obstructive sleep apnea, conductive hearing loss, obesity and spinal stenosis [8].

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