

What was Quasimodo suffering from?

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ABSTRACT

One of the fictional characters in Victor Hugo's novel "Notre-Dame de Paris" is without doubt Quasimodo: the hunchbacked bell-ringer of the Cathedral. From a medical perspective, the affliction of Quasimodo has remained a mystery for two centuries. Recent research however has linked his condition to a particular pathogenicity. Specifically, some passages in Hugo's novel suggest that a form of mucopolysaccharidosis, a deformity associated with many congenital and hereditary changes affecting the skeletal system, could explain Quasimodo's somatometric characteristics. In the present study we support the above claim and we discuss a number of related issues in the point between medicine and literature.

KEY WORDS: Notre-Dame de Paris; Quasimodo; literature; mucopolysaccharidosis; neurofibromatosis

Introduction

Deformities and monstrous characters have always been an important element in art [1]. Books and films have for a long time depicted outcasts, like Captain Hook in "Peter Pan", Erik in the "Phantom of the Opera", and the "creature" in "Frankenstein". In this paper we focus on Quasimodo, the emblematic fictional character in Victor Hugo's novel "Notre-Dame de Paris". The aim of our investigation is firstly to examine Quasimodo's condition from a medical point of view but also to discuss the symbolic gravity of this character both in relation to Victor Hugo and the times in which this famous French novelist has lived.

A question arises at this point. Why do we start a discussion on Quasimodo? And in general,

what attracts people in ugliness? A possible psychological explanation is that the aesthetic capacity towards ugliness has been attributed to our early experience of the feminine-maternal [2]. According to a number of psychologists, beauty is experienced at a certain stage of our development as both awesome and awful. From a philosophical perspective Immanuel Kant [3] has given the explanation that a person can be attracted on an ugly object simply because of the expectation that a certain order and harmony will eventually be found in it. Umberto Eco, in his book "On Ugliness" [4], unfurls a taxonomy of stark visual images of violence, deformity, immorality and cruelty, as well as quotations from sources ranging from Plato to radical feminists.

Quasimodo probably was suffering by a muco-

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polysaccharidosis, although the literature is still suggesting that neurofibromatosis is the most possible cause of his deafness and skeletal deformities [5, 6].

Who was Quasimodo?

Quasimodo draws the attention of the reader from the first moment in Hugo's novel, with his awful appearance, distorted body and strange behavior (**Fig. 1**). Hugo's descriptions in the chapter "Quasimodo" of the first book are presented below [7]:

We shall not try to give the reader an idea of that tetrahedral nose, that horseshoe mouth; that little left eye obstructed with a red, bushy, bristling eyebrow, while the right eye disappeared entirely beneath an enormous wart; of those teeth in disarray, broken here and there, like the embattled parapet of a fortress; of that callous lip, upon which one of these teeth encroached, like the tusk of an elephant; of that forked chin; and above all, of the expression spread over the whole; of that mixture of malice, amazement, and sadness. Let the reader dream of this whole, if he can.

A few lines later, Hugo shares more information about his hero:

A huge head, bristling with red hair; between his shoulders an enormous hump, a counterpart perceptible in front; a system of thighs and legs so strangely astray that they could touch each other only at the knees, and, viewed from the front, resembled the crescents of two scythes joined by the handles; large feet, monstrous hands; and, with all this deformity, an indescribable and redoubtable air of vigor, agility, and courage, – strange exception to the eternal rule which wills that force as well as beauty shall be the result of harmony. ... One would have pronounced him a giant who had been broken and badly put together again.

Among other things Quasimodo is deaf. In the following dialogue we read:

"What a devil of a man!" said Robin Poussepain still all bruised with his fall. "He shows himself; he's a hunchback. He walks; he's bandy-legged. He looks at you; he's one-eyed. You speak to him; he's deaf. And what does this Polyphemus do with his tongue?"

"He speaks when he chooses," said the old woman;



Fig. 1: Lon Chaney as Quasimodo and Patsy Ruth Miller as Esmeralda in the 1923 film, *The Hunchback of Notre Dame*.

"he became deaf through ringing the bells. He is not dumb."

Hugo uses Quasimodo's character to promote specific views: the unstable female nature as in the case of Esmeralda, who is moved by the charms of young Phoebus while staying indifferent to the feelings of ugly Quasimodo; the hypocrisy of a society that accepts the rejection of the outcast by soothing its conscience with ploys and all this with the help of the clergy, as it is the case of the informal adoption of Quasimodo by the archdeacon Claude Frollo; the inability to overcome one's sexual urge because of the religious prejudices that he serves, as is the case of the archdeacon, whose silent passion leads him to crime and his tragic end.

With regards to the name that Hugo chooses for his character, it turns out that this is not at all unplanned. The name Quasimodo obeys to an unwritten law of the literature according to which names often serve the logic and foreshadow of the plot [8].

The day that Frollo discovered the newborn in a sack, was the Sunday after Easter, the one called in the Eastern Church tradition as "Thomas Sunday" and in the Western Roman Catholic Church

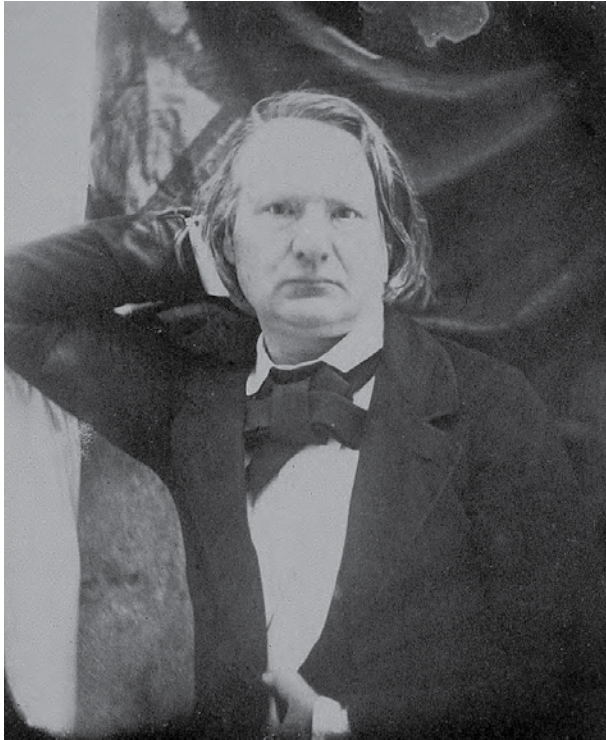


Fig. 2: Daguerreotype of Victor Hugo in 1853.

as “Divine Mercy Sunday” or “Octave of Easter” or “Quasimodo day”. The latter comes from the words of the introit in Latin: “Quasi modo geniti infantes, rationabile, sine dolo lac concupiscite”, which is translated as “as newborn babies, desire the rational milk without guile” [9]. We read in chapter “Claude Frollo” from the fourth book of the Hugo’s novel [7]:

He baptized his adopted child, and gave him the name of Quasimodo, either because he desired thereby to mark the day, when he had found him, or because he wished to designate by that name to what a degree the poor little creature was incomplete, and hardly sketched out. In fact, Quasimodo, blind, hunchbacked, knock-kneed, was only an “almost.”

In a footnote, Hugo gives another explanation for the choice: *The Latin word Quasimodo also means “almost” or “nearly”.*

Victor Hugo and his era

“Romanticism” is the intellectual movement in literacy and music that appeared in Europe in

the early 19th century, expressing an idealistic approach of the world as opposed to the rationalism of the Enlightenment. Victor Hugo (Fig. 2) was at the forefront of this movement. In his theatrical plays, novels and poems he described the struggles in the social life and the suffering of the lower classes. But Hugo’s biggest contribution to Romanticism is his firm interest for universal values. With the range and power of his pen Hugo criticized the injustices that were produced by the ruling classes at a time when it was difficult for anyone to understand injustices let only to fight against them [10]. Rightfully so, Hugo is considered today as a classic representative of liberalism in literature.

Hugo decides to write “Notre-Dame de Paris” while still studying at the Paris School of Philosophy [11]. The novel comes out in 1831, when Hugo was 29 years old and is a success and translated in other languages. On the pretext of a love affair between Esmeralda and Phoebus de Châteaupers, the unfulfilled libido of archdeacon Frollo that results in his deadly jealousy, but also the emblematic figure of Quasimodo slipping among the other characters in the drama, the author grasps the opportunity to beat the drum of the need to protect the medieval cathedrals that are about to collapse, something that in fact was the subject of doctorate thesis at that times. In Notre Dame, Hugo finds the perfect example in order to pass his positions to a society that turns a deaf ear to his words of warning about the decay of these buildings. It is in one such building that his hero has grew up and it is there where he has been deaf supposedly by the sound of the bells.

For the unsuspecting traveler who decides to visit Paris, Notre Dame is not only an unparalleled attraction, but summarizes in its niches and pediments the local history from the Middle Ages to the present day (Fig. 3). This metropolitan temple stands majestically on the isle of the River Seine, the famous Ile de la Cité. Its construction began officially in 1163 at the initiative of the Paris bishop Maurice de Sully, but its basic form was only completed in the 13th century [12]. During



Fig. 3: Notre Dame of Paris. Towers on west facade.

the 17th century, but mainly during the difficult years of the French Revolution, it suffered serious damage that led to frequent repairs and repeated reconstructions. In the mid-19th century, Eugène Viollet-le-Duc underwent a radical restoration. It is quite remarkable that the restoration work took place in the quarter century 1845-1870, immediately after the publication of Hugo's novel, practically giving affirmative response to his calls to rescue to Gothic's medieval cathedrals that were progressively collapsing [13-15]. In the third book of the novel [7], in the chapter titled "Notre-Dame", we read:

Thus, to sum up the points which we have just indicated, three sorts of ravages to-day disfigure Gothic architecture. Wrinkles and warts on the epidermis; this is the work of time. Deeds of violence, brutalities, contusions, fractures; this is the work of the revolutions from Luther to Mirabeau. Mutilations, amputations, dislocation of the joints, "restorations"; this is

the Greek, Roman, and barbarian work of professors according to Vitruvius and Vignole.

From the turbulent history of the cathedral, it is worth mentioning the coronation of King Henry VI of England on December 16, 1431, during the Hundred Years' War, as well as the brilliant Te Deum in the Choir of the cathedral on the reign of Louis XIV in 1669. At Notre Dame was also celebrated the Cult of Reason at the time of the reestablishment in 1793. And there was the coronation of Napoleon I on December 2, 1804. On August 26, 1944, with the withdrawal of occupying German troops from the city, a Mass is being attended to celebrate the liberation of Paris [16].

Notre Dame in Hugo's work is the point of reference for the episodes that evolve on many different levels: the geographical common place where the heroes' fates meet, the canvas where the plot of the drama unfolds. It is there where the adopted Quasimodo grows up, and it is there where he is supposedly deafened by the toll of the bell. Esmeralda escapes in the crypt of the church. Notre Dame is surrounded by beggars from the Court of Miracles with Clopin Trouillefou as their King (the King of Thunes) seeking to release the beautiful gypsy from the hands of archdeacon Claude Frollo. Notre Dame is also besieged by King Louis' troops to capture Esmeralda and hang her.

Mucopolysaccharidosis or neurofibromatosis?

The classic description of neurofibromatosis has been published by von Recklinghausen in 1882, while he was professor of pathology in Strasbourg, 51 years after the work of Victor Hugo [17]. Nevertheless, the disease had been partially described in 1783 by Tilesius [18]. According to Cox [5], the affliction that Hugo imposes on his character (extensive soft tissue and skeletal deformity with the preservation of motor skills and normal higher mental function) may be seen in severely affected cases of neurofibromatosis. The wart over Quasimodo's right eye could well have been a cutaneous neurofibroma affecting the eyelid. His spinal scoliosis, distorted thighs and legs, large hands and feet, club foot, huge head, and

irregular teeth are also seen in neurofibromatosis when central lesions of bone result from expansive growth of neurofibromas within the medullary cavity.

About forty years after the publication of von Recklinghausen, Gertrud Hurler, a German pediatrician, described a strange syndrome that is characterized by corneal clouding, skeletal abnormalities, and mental retardation [19]. Two years ago, during World War I, Charles Hunter had described a similar disease [20]. Hurler did not mention Hunter's paper because of interrupted communications caused by the war. Nowadays, due to the evolution of molecular biology, we know the pathogenesis of both diseases. They belong to the great family of "mucopolysaccharidoses".

Mucopolysaccharidoses are hereditary, progressive diseases caused by mutations of genes coding for lysosomal enzymes needed to degrade glycosaminoglycans. The major glycosaminoglycans are chondroitin-4-sulfate, chondroitin-6-sulfate, heparin sulfate, dermatan sulfate, keratan sulfate, and hyaluronan. These substances are synthesized and linked to proteins to form proteoglycans, major constituents of the ground substance of connective tissue, of nuclear and cell membranes [21]. The hyaluronan is an exception in this mechanism.

Failure of degradation because of absent or grossly reduced activity of mutated lysosomal enzymes results in the intralysosomal accumulation of glycosaminoglycan fragments. The characteristic pattern of clinical, radiologic, and biochemical abnormalities are due to cell dysfunction caused by the accumulation of distended lysosomes in the cells [22].

Mucopolysaccharidosis I (MPS-I), or Hurler syndrome, is caused by mutations of the *IDUA* gene on chromosome 4p16.3 encoding α -L-iduronidase. Deficiency of α -L-iduronidase results in a wide range of clinical involvement, which are ends of a broad clinical spectrum. Hurler disease is a severe, progressive disorder with multiple organ and tissue involvement. Its

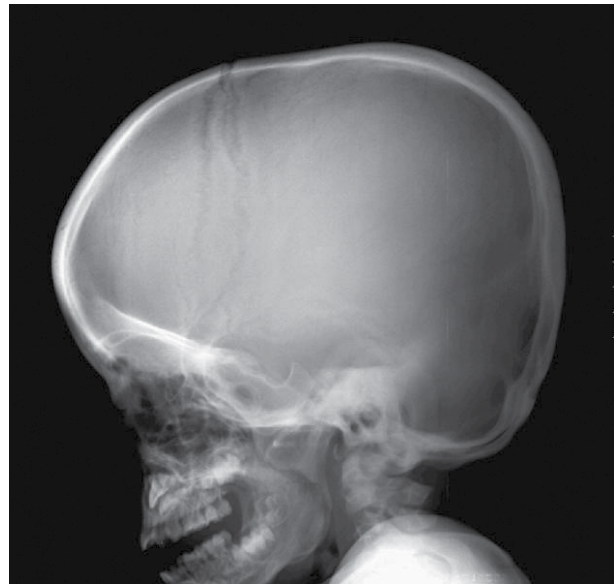


Fig. 4: In Hurler syndrome, the head can be large with prominent frontal bones and the skull can be elongated.

transmission is autosomal recessive. Diagnosis is usually made between 6 and 24 months with evidence of hepatosplenomegaly, coarse facial features, corneal clouding, large tongue, prominent forehead, joint stiffness, short stature, and skeletal dysplasia [23]. Facial deformities are usually pathognomonic (**Fig. 4**): frontal bossing, prominent eyes with hypertelorism and depressed nasal bridge, gapped teeth, gingival hypertrophy and (as it has already been described) a thickened tongue.

Facial features resemble the gargoyles, the carved or formed grotesque with a spout designed to convey water from a roof and away from the side of a building, thereby preventing rainwater from running down masonry walls and eroding the mortar between. [24]. Architects often used multiple gargoyles on a building to divide the flow of rainwater off the roof to minimize the potential damage from a rainstorm, like Eugène Viollet-le Duc in Notre-Dame. Hunter used the term "gargoylism" in order to describe his own cases (**Fig. 5**), but nowadays the term is used for Hurler's disease.

Most patients have recurrent upper respiratory



Fig. 5: A gargoyle on the tower of Notre Dame of Paris.

ry tract and ear infections, noisy breathing, and persistent copious nasal discharge. Most children with Hurler syndrome acquire only limited language skills because of developmental delay, combined conductive and neurosensory hearing loss, and enlarged tongue. Progressive ventricular enlargement with increased intracranial pressure caused by communicating hydrocephalus also occurs. Corneal clouding, glaucoma, and retinal degeneration are common. Radiographs show a characteristic skeletal dysplasia (**Fig. 6a**). The earliest radiographic signs are thick ribs and ovoid vertebral bodies. The long bones have enlarged, coarsely trabeculated diaphysis with irregular metaphyses and epiphyses. With progression of the disease macrocephaly develops with thickened calvarium, premature closure of lambdoid and sagittal sutures, shallow orbits, enlarged J-shaped sella, and abnormal spacing of teeth with dentigerous cyst.

The body's height remains short with kyphoscoliosis at thoracolumbar spine. Mental retardation also exists. In a broad pelvis there are dysplastic acetabuli. The femoral necks are in valgus position. The appearance of ossific nucleus of the

femoral heads retards. A subluxation or true dislocation of both hips is also exists (**Fig. 6b**). These patients die early in life because of cardiac insufficiency or respiratory infections.

Hunter disease, or mucopolysaccharidosis II (MPS-II), is a linked disorder caused by the deficiency of iduronate 2-sulfatase. As an X-linked recessive disorder, Hunter disease manifests almost exclusively in males. Marked molecular heterogeneity explains the wide clinical spectrum of the disease. Patients with severe MPS-II have features similar to those of Hurler disease except for the lack of corneal clouding and slower progression of somatic and central nervous system deterioration. Coarse facial features, short stature, dysostosis multiplex, joint stiffness, and intellectual disability manifest between 2 and 4 years of age. Extensive, slowly progressive neurologic involvement precedes death, which usually occurs between 10 and 15 years of age [20].

Patients with the mild form can have a near-normal or normal life span, minimal central nervous system involvement and slow progression of somatic deterioration with preservation of cognitive function in adult life. Survival to ages 65 and

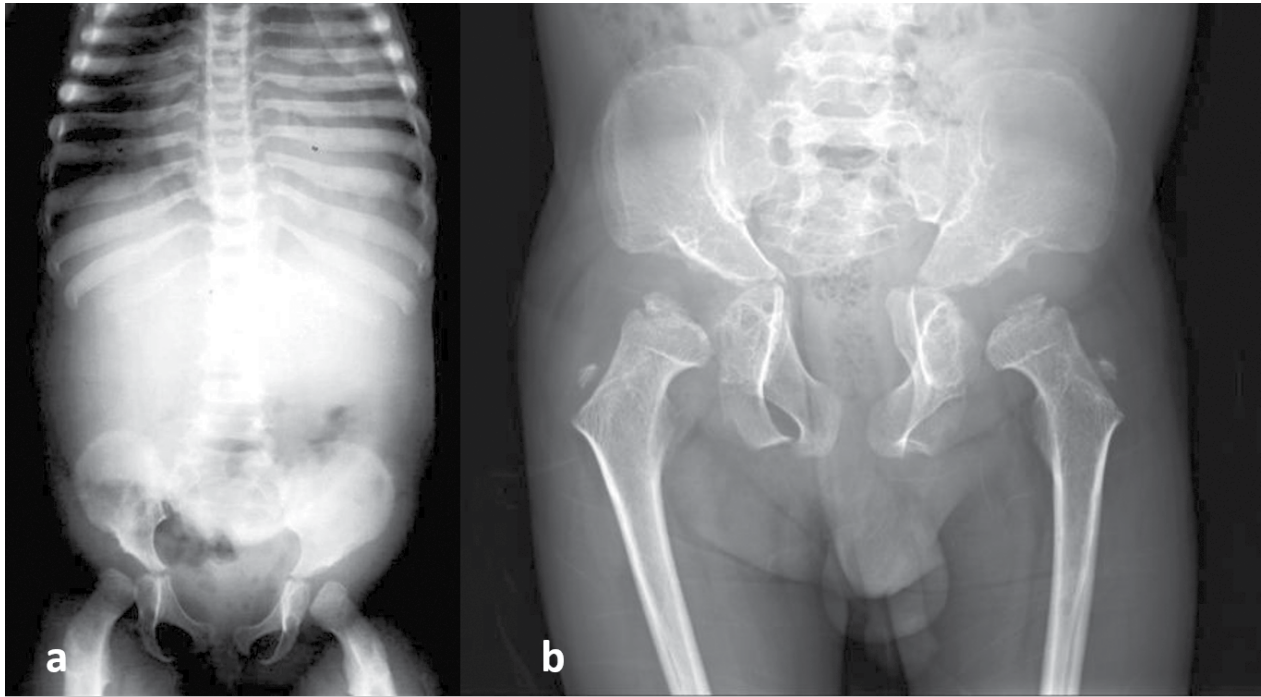


Fig. 6: Kyphoscoliosis of thoracolumbar spine (a), and subluxation of both hips (b) in a patient suffering by Hurler syndrome.

87 years has been reported. Somatic features are Hurler-like but milder with a greatly reduced rate of progression. Adult height may exceed 150 cm. Airway involvement, valvular cardiac disease, hearing impairment, carpal tunnel syndrome, and joint stiffness are common and can result in significant loss of function in both the mild and severe forms.

Discussion based upon a hypothesis

Hugo's work gives a believable description of the Quasimodo's looks and this could well fit in to Hurler's description. However, the absence of an obvious mental disability in Hugo's hero makes it difficult to come to reliable conclusions with regards the above statement and we surely accept that it is Hugo's privilege to form a character according to his imagination, perceptions, and foreshadowing. Under these circumstances and beyond his somatometric measurements, Quasimodo, as a fictional character, ought to have the ability to understand the events in his surround-

ings; the will to think and act with efficiency; the sensibility to feel and to love.

It is a common secret that parthenogenesis does not exist, both in nature and the creative mind of an author. From this point of view, it would be utopian to suggest the opposite and, as in the case of Notre Dame, it is very likely that Hugo's perception of a known person with similar characteristics stirred him to design his own hero [26]. Based on this assumption, the center of gravity of our discussion should be shifted from Quasimodo himself to the man who apparently inspired his creator. Hugo in a particularly revealing apostrophe writes in the chapter "Three human hearts differently constructed" of the eight book of the novel [7]:

No one had yet seen in the gallery of the statues of the kings, carved directly over the arches of the portal, and strangled the spectator, who had, up to that time, observed everything with such impassiveness, with a neck so strained and visage so hideous that, in his motley accoutrement of red and violet, he might have

been taken for one of those stone monsters through whose mouths the long gutters of the cathedral have discharged their waters for six hundred years.

Quasimodo's resemblance to gargoyles, the grotesque demonic figures that project from buildings, is obvious. Gargoyles supposedly removed the satanic forces. From a practical point of view however, they just conveyed water from a roof of a building away from its sides. Gargoyles were in fact artful gutters and were proposed by Eugène Viollet-le-Duc himself [24].


In August 2010, Adrian Glew published an article about the "true life of Quasimodo," a lithographer working in Notre Dame in 1820. The story taken down by Glew can be found in the memoirs of Henry Sibson, a sculptor from England who worked at Notre Dame at the same time that Hugo wrote his novel [27]. Sibson describes the sculptor as hunchback and deformed giant, although his name misses him. As Victor Hugo supervised the restoration work of the cathedral, it is quite possible that he aware of the existence of this man who went by the name "Le Bossu" ("the hunchback" in French). His real name was Monsieur Trajin, and Adrian Glew reveals that Hugo and Le Bossu lived in the same town of Saint Germain-des-Pres in 1833. Apparently Hugo used this man's name,

transforming it from Jean Trajin to Jean Valjean to create the central hero in his masterpiece "Les Misérables".

Epilogue

Regardless of whether the real Quasimodo existed and irrespectively of what his real affliction was, our fictional hero is a universal symbol of resistance with his "indescribable and redoubtable air of vigor, agility, and courage", according to Eco [4].

Quasimodo is a "strange exception to the eternal rule which wills that force as well as beauty shall be the result of harmony" [7]. It is common place that there has been discrimination against the disabled people and the people with unusual bodies in a number of fields, like employment, voting, marriage and parental rights, the right to travel, the right to be free from institutionalization, and the right to have access to the courts of law.

In our opinion, Quasimodo symbolizes our fellow man; our fellow "other" who overcomes social and ideological obstacles. Quasimodo hopes, loves and fights. 

Conflict of interest:

The authors declared no conflicts of interest

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