

Modern Verse, but one should be chary of dismissing Yeats's poetic judgment. One of Gogarty's poems ends like this:

"Beauty never comes on earth
But an equal Grief is born;
Hidden, maybe, in the dearth
Of the hours ere the morn;
Or that in her core are strife,
Gain of Love and loss of Life.

This is nothing new at all;
We have heard it all before;
Beauty one side of the Wall,
On the other side, the War.
Love and Death; and no denying
These things do not end by dying."

James Stephens told Gogarty that it was unlikely that he would be recognised as a poet as he should be until 50 years after he was dead. There are still 27 years to go. When young he had a great facility for light verse and parody, produced in profusion, scattered through his letters, and published wherever he could use them to raise some money. That he did not achieve more praise for his later work may be due to the oddity that people who become famous for their wit seldom acquire the highest regard of their contemporaries whether in art, administration, or scholarship. Sidney Smith asserted that he had sunk through his own levity whereas his theologian brother had risen by his gravity.

Those Gogarty sayings that stick in the mind have a little Irish twist to them such as, "There is nothing more sinister than the

humility of the mean." Of Sir Thornley-Stoker he said that he "always spoke of his fee as his 'honorarium' for who can pay a doctor all he owes?" Of Browning's obscurities Gogarty wrote, "When I read his translation of Aeschylus I find it very useful to have the Greek beside me so that I may find out what the English means." His comment on a man coming out of a pub consulting a compass (where else but in Dublin?) is typical, "He goes home by compass! He cannot trust the ebbing town." Let me close with one last quotation from this unusual surgeon whose prose as well as whose poetry is not as well known as it deserves to be. It takes an Irishman to have written:

"To be as magical as Life itself, and as irresponsible; to be lunatic enough to take the hard edge from knowledge, to be irrational enough to temper Justice with Mercy, and to be able to adjust oneself to the changes in intensity which the waxings and wanings of reality assume in the shimmerings of its cloud—this would be an ideal adjustment and a poetic opposition to outrageous Fortune."

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Quest for Quasimodo

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Perhaps the best known character in the whole of French literature is Quasimodo, the hunchback of Notre Dame. Even those who have never read Victor Hugo's book *Notre-Dame de Paris* must be familiar with the many famous cinematic portrayals of the character (fig 1). In his book Victor Hugo gives an unforgettable portrait of the misshapen bell ringer and his physical deformities: "that tetrahedral nose, that horseshoe mouth, of the tiny left eye, obstructed by a bushy red eyebrow, while the right eye had vanished entirely beneath an enormous wen, of those irregular teeth, notched here and there like castle battlements, of that horny lip on which a tooth encroached like an elephant's tusk, of that cleft chin . . . a huge head sprouting red hair; between the two shoulders an enormous hump, the repercussions of which were evident from the front; a system of thighs and legs so strangely warped that they met only at the knees and looked, from the front, like two scythe blades joined at the handle; broad feet and monstrous hands; and somehow, along with such deformity, an appearance of formidable energy, agility and courage."¹ This was the portrait Hugo painted of Quasimodo at the age of 19, during the election of the fools' pope. Elsewhere in the book he gives a description of Quasimodo as a 4 year old child: "a sort of hideous little monster, lame, one-eyed and misshapen, was dragging itself across the floor squealing."² Finally Victor Hugo describes his skeleton: "they noticed that its spinal column was

curved, that its head was between the shoulder blades and that one leg was shorter than the other."³

Von Recklinghausen's neurofibromatosis

Although *Notre-Dame de Paris* is a work of fiction, this careful and extensive description of both soft tissue and skeletal abnormalities

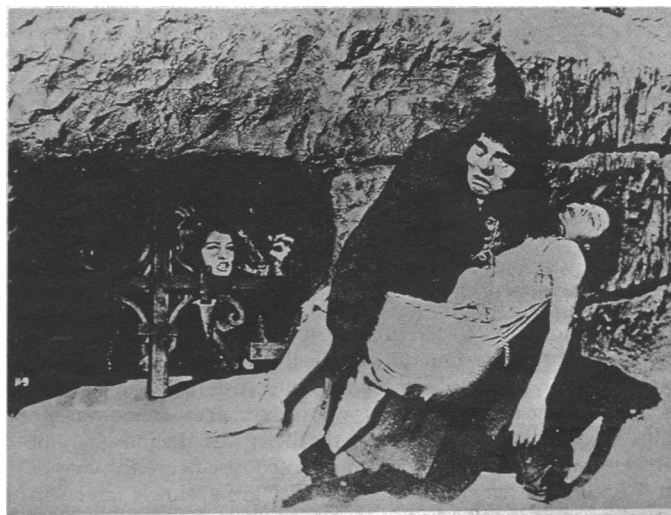


FIG 1—Lon Chaney as Quasimodo in silent film in 1923 of *The Hunchback of Notre Dame*.

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can be recognised as belonging to a single medical entity. The affliction that Hugo imposes on his character first occurs in early childhood and combines extensive soft tissue and skeletal deformity with the preservation of motor skills, including strength, agility, and normal higher mental function. Such a combination of abnormalities may be seen in severely affected cases of Von Recklinghausen's neurofibromatosis. If you go through his description of



FIG 2—Cover of 1831 edition of *Notre Dame de Paris* by Victor Hugo, showing Quasimodo and Esmeralda.

Quasimodo with this disorder in mind the deformities that he lists are consistent with this diagnosis. For example, the wart or “wen” over his right eye could well have been a cutaneous neurofibroma affecting the eyelid. His skeletal anomalies—that is, spinal scoliosis, distorted thighs and legs, large hands and feet, club foot, huge head, and irregular teeth—are all seen in neurofibromatosis when central “cystic” lesions of bone result from expansive growth of neurofibromas within the medullary cavity.⁴ In Von Recklinghausen's neurofibromatosis “kyphoscoliosis and pseudoarthroses (especially with bowing of the tibia and fibula) are common . . . overgrowth of the cranial bones . . . macrocephaly and a variety of other anomalies may be observed, including elevated scapulas and club foot . . . and involvement of the bony maxilla and mandible.”⁴

Effects and origins of the disease

The condition is a well recognised cause of bilateral deafness and Hugo records that Quasimodo went deaf at the age of 14 after his appointment as a carrionist at Notre Dame. Although Hugo makes Quasimodo allege that his deafness was of sudden onset and was the result of exposure to the noise of the bells, the diagnosis is not entirely inconsistent with this. Although acoustic neurofibromas of Von Recklinghausen's disease normally grow slowly and “in classical descriptions the deafness is of insidious onset and gradual progression, in many instances in practice, the onset is of sudden

deafness.”⁵ Deafness may precede other central manifestations by many years. Hugo's supposition that Quasimodo already suffered from skeletal deformities at the age of 4 is likewise consistent with the diagnosis, as in 60% of cases the disease is evident by the age of 2.⁶

Victor Hugo's description was written 51 years before the classic description of the disease in 1882 by Von Recklinghausen while he was professor of pathology in Strasbourg;⁷ *Notre-Dame de Paris* was first published in Paris in 1831 (fig 2). Nevertheless, the disease had been partially described before this time. Tilesius described the cutaneous lesions in 1783,⁸ and Smith in 1849 described other manifestations but failed to appreciate their nerve sheath origin.⁹

A similar portrayal of the horrific cutaneous and skeletal disorders that may occur in the disease was given by Sir Frederick Treves in 1885, describing the elephant man of Whitechapel (fig 3).¹⁰ The drawings made of this unfortunate creature at that time could well be used as illustrations for Hugo's masterpiece.

Victor Hugo's portrayal of Quasimodo gives an excellent and remarkably detailed description of the disabilities imposed by Von Recklinghausen's disease. The clarity and clinical consistency of the description leaves little room for other diagnoses and would suggest that it was based on a living being that Hugo either knew or had met. There is unfortunately no suggestion in his works or in the writings on Hugo by others of a possible identity or origin for this unforgettable creation. The only suggestion hitherto put forward is that Quasimodo was a refinement of previous characters, but a more detailed examination of these earlier “models” for Quasimodo does not evoke the same immediate recognition of a consistent clinical entity. Edwards suggests that Habibrah, the twisted ugly dwarf in *Bug-Jargal*, later appeared with appropriate changes as Quasimodo in *Notre-Dame de Paris*.¹¹ *Bug-Jargal* is a little known work by Victor Hugo first published in 1819 and is the story of a negro revolution against European rule. Pringle has pursued a similar theme: “Quasimodo had been foreshadowed in the dwarf Habibrah of *Bug-Jargal* and *Han d'Island*, but these were merely sketches, Quasimodo is an immortal and perfect picture.”¹² *Han d'Island* (*The Outlaw of Iceland*) is another of Victor Hugo's early works first published in 1823. In *Bug-Jargal* Victor Hugo writes: “Habibrah was not a negro, nor yet, to speak correctly a mulatto. As to stature



FIG 3—John Merrick, elephant man, in 1890.

he was literally, a dwarf, and his figure, on the whole, was so deformed, that a stranger could scarcely look at him without shuddering... his body was short but of amazing bulk and was supported on limbs which, to judge from appearances, were quite inadequate for their burden. To the huge head which reposed on his short neck."¹³ Han d'Island is described thus: "A little man short and thick set... his beard was red and bushy, and his head... seemed to be thickly covered with hair of a similar colour, his mouth was large, with very thick lips; his teeth white and pointed with wide spaces between them, his nose was curved like the beak of an eagle, his grey blue eyes."¹⁴ Neither of these descriptions shows any clear, clinically recognisable details, and Quasimodo bears little physical resemblance to either; the common thread being only one of deformity. Hugo himself gives no clue in his autobiographical writings to his model for Quasimodo. No recognisable model exists in his drawings, nor is the subject mentioned in the many biographies and studies of his life and work.

Did Quasimodo ever live?

Victor Hugo was only 29 when *Notre-Dame de Paris* was published in 1831, but he had begun to consider the book as early as 1828. Until that time he had always lived in Paris, except for brief stays both in Italy and Spain with his military father. Although Hugo may have encountered some such deformed victim of Von Recklinghausen's disease while on these foreign visits, it is more likely that he encountered his model in Paris. Had such a person held any prominent position in the public life of the city when the book was published, however, he would probably have become as

notorious in his time as the elephant man was many years later. The model that Hugo used for his portrayal might have been already dead when the book was published, and any person with deformities of the severity that Hugo attributes to his central character would probably have a much reduced life expectancy. Although the mystery of who it was that Hugo portrayed remains unsolved, the portrait that he made so unforgettable has incontrovertibly conferred upon the unfortunate subject, whoever he was, a posthumous immortality.

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Unsuitable reading?

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BMA House, designed by Sir Edwin Lutyens and built on the site of a house where Charles Dickens lived in the mid-1800s, is—as you all know—located at Tavistock Square in London. In the same location, if you look carefully, you will also find the Cadaver Club, a typical British institution founded by men for men—with an interest in murder. The club is exclusively masculine, women are neither admitted as members nor entertained. The committee takes the view that their presence would hardly compensate for the expense of putting in a second set of lavatories. But books do not need toilets and even if lady writers are excluded their books are not—they are fairly well represented on the shelves of the club library.

Well, the Cadaver Club does not exist in reality, only in the third book by P D James, *Unnatural Causes*. The male members might give expense as their reason for excluding women—personally, I believe that their real reason was that they feared competition or even defeat. So many of the best crime writers of bygone and present days have been or are women that it should send cold chills up the backs of the male committee members. And who can give a good explanation for the high percentage of ladies in this morbid genre? Do ladies have more imagination than men, can they construct better plots? Or is murder their way of fighting for women's liberation? I do not have an answer, but the list of lady writers is

indeed impressive: Dorothy L Sayers, Agatha Christie, Margery Allingham, Ngaio Marsh, and P D James, to mention only a few of the most outstanding.

Today P D James is highly acclaimed and ranked not only as an equal to Dorothy L Sayers and Agatha Christie but as their superior. The main reason is that her characters are much more human, less stereotyped, much more alive—until they get murdered—than those of her famous predecessors, in short, that she is a better writer. Since 1962 nine novels by Mrs James have seen the light of day—or is the dark of night more appropriate for crime novels? All are exciting; *Innocent Blood* is a thriller, all the others classic detective stories, many set in an environment for which Mrs James's long experience as an administrator in Her Majesty's National Health Service has formed the background. She skilfully varies her settings but in most of the books keeps to the sociomedical sphere so well known to her. In her company we visit a nursing school (*Shroud for a Nightingale*) and a forensic science laboratory (*Death of an Expert Witness*), we are taken to a private home for the young disabled (*Black Tower*) and to a fashionable psychiatric clinic (*A Mind to Murder*). In the vicinity of an Elizabethan manor house (*Cover Her Face*), the tranquility of which is shaken by murder, we find St Mary's Refuge, a home for unmarried mothers, and in *Innocent Blood* we follow Philippa Palfrey into the visitor's room of Melcombe Grange, a prison.

Mrs James has realised that there is hardly any murder method that will escape detection with today's modern scientific techniques—if foul play is at all suspected—and in most cases lets her murderers, among them doctors and nurses, kill by simple and well established methods: the blunt instrument or the murderer's own hands. She has a fancy for strangulation and suffocation—she says

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