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Fragile (2005): A case of osteogenesis imperfecta

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Summary

When Mercy Falls hospital is about to close its doors for good, a series of strange phenomena begin to take place: the children start to suffer fractures for no apparent reason. A nurse, Amy, suspects that these occurrences are related to a little girl called Mandy who had been a patient in the hospital years before and who had the same symptoms as the children now suffering from fractures: a tendency to bone fractures owing to what is known as brittle-bone disease. Amy then begins to investigate whether there is some kind of relation between Mandy's case and those of the children in her care.

Keywords: Osteogenesis Imperfecta, Genetic Disease, Brittle Bones, Treatment, Evolution.

Technical details

Title: *Fragile*
Original Title: *Frágiles*
Country: Spain and UK
Year: 2005
Director: Jaume Balagueró
Music: Roque Baños
Screenwriter: Jaume Balagueró and Jordi Galcerán.
Cast: Calista Flockhart, Richard Foxburgh, Elena Anaya, Gemma Jones, Colin McFarlane, Michael Pennington, Daniel Ortiz, Yasmin Murphy, Karmita Cervera and Ivana Baquero.
Color: Color
Runtime: 104 minutes
Genre: Horror
Production Companies: Castelao Productions, Just Films and Future Films
Synopsis: An old paediatric hospital on the Isle of Wight (Great Britain), about to close its doors, is haunted by a little girl who died more than forty years before under strange circumstances.
Awards and nominations: Goya (2006) for the best special effects. Nominated for the best original music.

Osteogenesis imperfecta, or brittle bone disease, belongs to the category of rare diseases[1] and not many films have used it as background. The exceptions are *Unbreakable* (2000), by M. Night Shyamalan, in which the main character suffers from type I, *Amelie/ Le fabuleux destin d'Amélie Poulain* (2001), by Jean-Pierre Jeunet, where a supporting character has it, *Sixth Happiness* (1997), by Waris Hussein, in which a patient with this pathology is the main character, and *Fragile*, the subject of the present analysis.

The film

Mercy Falls hospital for children is about to close its doors after more than a century of service. Most of the staff has been dismissed and the installations have practically been dismantled. Amy (Calista Flockhart), an American nurse, arrives at Mercy Falls to take over the night shift while the last children to remain are being taken to other hospitals. But very soon Amy detects something strange about the place. Maggie (Yasmin Murphy), one of the little girls at the hospital, is nervous, frightened, and talking about something intangible, invisible; something that shouldn't exist, something full of pain and hate - something she calls the "mechanical girl". She lives upstairs, on the second floor, the floor that was closed almost forty years before. Little by little this presence makes itself more and more evident, more aggressive, moving about the hospital corridors, hiding in the

shadows. As it turns out, the hospital is hiding a secret, and, to discover the truth and protect the children, Amy will have to face up to those who do not believe her, to her own fears, her own past and the truth[2].

Figure 1: X-ray of the fracture in one of the legs of a child at the hospital

But this truth is much darker than what anybody suspects. This truth has a name, Charlotte (Karmeta Cervera), a nurse who worked at Mercy Falls forty years before and who had an obscure past. She was obsessed with one of the girls in the hospital, Mandy (Ivana Baquero), who suffered from osteogenesis imperfecta, and caused her to have fractures so that she would not recover and be transferred from the hospital. One day Charlotte smothered Mandy and then committed suicide after putting the girl's metallic braces on.

However, her spirit never left and remained there next to what she most needed: the children. Thus when Mercy Falls is getting ready to close and is evacuating the children, Charlotte appears to try to put a stop to it. To do this, she acts on the children by causing fractures proper to brittle bone disease, which is why the children begin to have fractures with no apparent cause (figures 1, 2 and 3).

Osteogenesis imperfecta

Osteogenesis imperfecta, commonly known as "brittle bone disease", is a congenital disease involving abnormal fragility of the bones without any apparent cause.

It is caused by a genetic defect that affects the production of collagen, which is the main protein of connective tissue. A person who has this disease has a lower amount or poorer quality of collagen than normal, making the bones extremely fragile and brittle. For example, one could break a bone in the leg just by sneezing or turning over in bed while sleeping.

It is sometimes possible to establish a diagnosis based simply on clinical data. In certain cases there are biochemical (collagen) and molecular (DNA) tests available to help confirm the diagnosis. It usually takes several weeks to do these tests and in approximately 10 to 15% of the mild cases, the collagen test is negative even though the disease is present while 5% must be subjected to a genetic test.

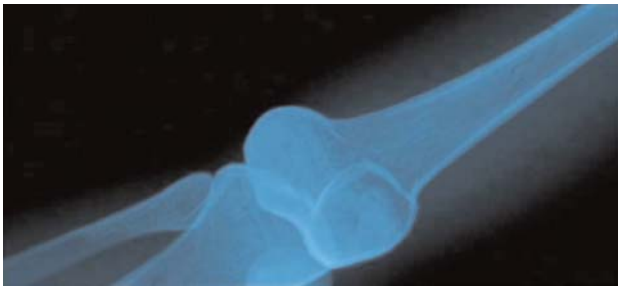

Figure 2: X-ray of the fracture in one of the legs of a child at the time when it is broken with no apparent cause

The symptoms vary widely from one individual to another, even among patients with the same type and of the same family and not all the symptoms are evident in each case. There are four types of osteogenesis imperfecta:

Type I: this is the mildest and most common type of the disease. Fractures tend to occur before puberty. Symptoms include normal or near normal stature, loose joints, muscle weakness, sclera with a blue, purple or grey tint, rectangular face, slight bone deformity, hearing loss beginning in the early 20s or 30s, normal collagen structure, but reduced amounts of it, and brittle teeth.

Figure 3: A nurse has a broken nose, with no apparent cause

Type II: this is the most severe form. It is frequently lethal shortly after birth, often owing to respiratory problems, although recently some patients with this type have reached adulthood. The symptoms include a predisposition to numerous fractures and severe bone deformity, small stature, underdeveloped lungs, loose joints, and collagen is improperly formed.

Type III: in this type, bone fractures are common normally as from birth, and X-rays reveal healed fractures that occurred even prior to birth. Symptoms include: lower stature than normal, blue, purple or grey sclera, loose joints and poor muscle development in arms and legs, triangular face, possible respiratory problems, often severe bone deformity, a barrel-shaped rib cage, possible brittle teeth, possible hearing loss, and improperly formed collagen.

Type IV: this type falls between Type I and Type III in severity. Bones fracture easily, especially before puberty. Stature is lower than normal, sclera are white or near-white (i.e. normal in colour), bone deformity is mild to moderate and there is a tendency to spinal curvature. Other symptoms are a barrel-shaped rib cage, triangular face, possibly brittle teeth, hearing loss, and improperly formed collagen.

Other symptoms of osteogenesis imperfecta are deformities in the arms and legs and hands and feet, ciphosis, a low nasal bridge, pectus excavatum (sunken or funnel chest), flat feet, hypermobility, a tendency to bruising, and bowed legs.

Since a cure has not yet been found for this disease, treatment is directed toward preventing or controlling symptoms, maximizing independent mobility and developing optimal bone mass and muscle strength. Likewise, care of fractures, extensive surgical and dental procedures and physical therapy are also recommended. The use of wheelchairs, braces and other mobility aids is common, particularly with the more severe types of the disease.

A surgical procedure called "rodding" is frequently used. This treatment involves the insertion of metal rods through the length of the long bones to strengthen them and to prevent or correct deformities.

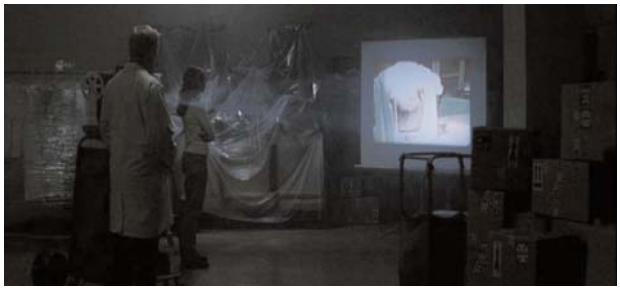

Figure 4: The doctor, on seeing Mandy's spine, explains to Amy that it is a case of osteogenesis imperfecta

Currently, the use of biphosphonates is being investigated, with some promising results in children with the disease. Other therapies include bone marrow transplant, the use of growth hormones and gene therapy. Finally, good nutrition and supervised exercise are key to optimizing bone and muscle strength [3-5].

Aspects of the disease that appear in the film

The first scene of the film shows a child sleeping in a hospital bed who suddenly, after hearing a series of strange noises, has a fracture in one of his legs with no apparent cause. When the doctor sees him, he presents an open fracture in the leg apparently caused by a brusque movement made during sleep, although the cause is not clear. Subsequently, while he is undergoing a CAT scan, another fracture occurs in the same leg, but this time it can be seen that the child has been completely still, making the case even more mysterious (figures 1 and 2). These are fractures typical of "brittle bone disease", but the strange thing is that the child had previously showed no symptoms whatsoever of this disease.

Figure 5: Video of Mandy's operation

However, the children in the hospital repeatedly mention that the cause of these strange phenomena is "the mechanical girl". Nurse Amy is dismayed by this name and begins to investigate what is going on. Thus, thanks to some clues that the "mechanical girl" gives her, she finds out that years before there was a girl hospitalized there named Mandy. One day Amy decides to go up to the closed-off upper floor to Mandy's room and there she discovers diverse instruments, including a wheel chair, typical of a person suffering from bone disease who cannot get around on their own These instruments, in particular the metal braces, provide Amy with the clue she needs to uncover the meaning of the "mechanical girl".

Later, in the scene where Amy and the doctor are viewing a tape found in Mandy's old room, both of them realize that the disease the girl had was osteogenesis imperfecta (figure 4).

This is the only scene which gives a description of the disease, albeit quite a brief one; in fact, several manifestations of it can be observed: the girl has curvature of the spine and bluish-coloured sclera, as well as deformities of the legs. It can also be glimpsed that the child is constantly bad tempered, although this is not a classic symptom of the disease.

Further on we see the treatments the girl has received: rehabilitation and physical therapy, as well as surgery, possibly to implant metal rods along her bones, since later on we can see that Mandy indeed has a series of metal instruments on her legs that allow her greater mobility (figures 5, 6 and 7).

Figure 6: Inserting metal rods

Although the film does not clearly state the type of osteogenesis imperfecta the girl was suffering from, it does insinuate that she was beginning to improve in response to the treatment received before dying. We can therefore directly rule out that she was suffering from Type II, which is usually fatal in most cases in early childhood. It could be Type III or IV, since hers is a severe form with progressive deformity. In fact, Mandy's symptoms are the most common ones of the disease and the "film within the film" does not show any specific symptoms that would elucidate the disease type.

Mandy's disease leads her to undergo important changes in her social surroundings: she must go

Figure 7: Metal rods on the leg

Figure 8: Charlotte, the nurse, with Mandy. It is possible to see the metal prostheses on the girl's arms

into Mercy Falls hospital, she suffers from strong changeable moods with bouts of anger, and she must depend on Charlotte. Charlotte is the nurse who, after suffocating Mandy and putting on her metal braces, decides to stop the children from leaving the hospital (figures 8 and 9).

Finally, there is another key element representative of the disease which is present throughout the film and even the preview: frequent scenes of bone x-rays in which a bone is cleanly broken with great ease, thus reflecting yet another aspect of the disease in question.

Figure 9: Charlotte, with the prostheses

Conclusion

Although the underlying idea of this film is quite original, the screenplay is somewhat banal and superficial, considering that the topic could have given rise to a more profound one. Furthermore, the aspects inherent to the disease are not made clear enough, leaving laypersons in the dark with unresolved gaps and questions owing to the amalgam of strange devices whose purposes are not made clear. For example, the type of osteogenesis imperfecta is not clear, nor is the treatment the girl is subjected to, since the film only gives a brief and superficial explanation of the disease (figure 4). Since the entire film revolves around this illness, its symptoms and its consequences, it would not have been amiss to make certain aspects of it more explicit so that the audience could clearly understand the development of the story.

It would thus have been more interesting to go deeper into a description of the disease to clarify certain aspects which continue to be a mystery for the audience even after having seen the film.

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