



**"NOISE of the CREATION"**

Video Documentary – 62min

**SYNOPSIS**

This unique videofilm tries to give the viewer an insight into metabolic diseases, by presenting an image of the scientific research, as well the changes in daily life for patients and all those involved. It is geared especially towards younger viewers, 10yrs and up, since is mostly young people who are affected.

**TIMELINE**



A timeline covering the past century takes from a life in mental institutions for retarded people to young adolescents who can now live somewhat normal lives.

**The CELL**

A number of scenes inside a giant human CELL show in tangible and comprehensive ways some of the processes which go wrong in a real human cell.

In an entertaining and fascinating way 4 eminent professors in the fields of genetics and metabolic disorders will present each one a segment highlighting aspects such as genetic errors, lysosomal errors etc.



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## **Manga CARTOON**

Also featured in the film are 4 CARTOON segments in Japanese Manga style. This substory will play about 50 years in the future. This is done for two reasons: firstly to engage the young viewers in a “cool” fashion. Second, it should aid in provoking discussion and afterthought. Further it will help the viewer to create a reference to where we stand with science and therapies.



## **CHAPTERS**

The film consists of four chapters:

### **1. THE DISEASE**

This chapter will focus on the disease itself. A historical view of the early research and advances in understandings. In the cell the origin of errors in the DNA is explained.

### **2. THE CONSEQUENCES**

This chapter will emphasize the various ways these disorders present themselves. We will see different patients, from very ill, to people suffering minor inconveniences. We'll be introduced to patient groups such as BOKS which were instrumental in setting up CEMA (regional centers for inherited metabolic disorders), which in a unique way supports patients and parents.

### **3. THE THERAPIES**

In recent years therapies have become available, mostly as ERT (enzyme replacement therapy) These advances were made possible by the “orphan-drugsact” which way instrumental in many of the advances we see today. We'll see advances in technical and medical field. Also the advent of “special foods” , dietary products, sometimes even made for one single patient.

### **4. THE FUTURE**

What is being worked on? What kind of life will children have which before, never reached adolescent age? The psycho-social aspects. Political aspects such financial issues and the conditions for releasing new drugs on the market. Where lies hope, what is reality, where are we now?

## **PATIENTS**

Throughout the film we will follow a few patients in their daily activities, highlighting where these lives differ from everyday normal people. This will also put in perspective what the consequences on the immediate surroundings of the patients. Whole families' lives change sometimes quite drastically. As one example we will follow a child with a severe mitochondrial disorder as he's taken to a special care home, in order to allow the family some relief in their daily lives. In this fashion the psycho-social aspect will be shown as well.

## **CEMA's**

A fair amount of attention will be paid to CEMA's. These centers are based on a unique co-operation in a team consisting of a pediatrician, a geneticist, psychologist, dietician, and even administrative assistant. The whole team works in concert to help the patients and parents as much as possible. These centers are unique for Belgium and have come to existence under pressure from such groups as BOKS vzw, an initiator of this project as well.

## **THE INDUSTRY - "new player"**

Several interviews with representatives from pharmaceutical companies involved in rare diseases will give an overview of the drugs available and in the pipeline. We will shed light on the new possibilities, but also the hurdles which remain on the path a drug has to travel from lab to patient. Safety- and political issues are still important for people desperately waiting. Attention to available therapies and diet foods will also be given.

## **TARGET AUDIENCE**



The film will be made for a wide as possible audience. It will be very comprehensive and Accessible In the film 3D computer molecular animations and people in white coats explaining things are avoided. That's one of the reasons to choose for the CELL. This gives a much more involved and engaging picture of what goes on in a human cell. The cell will be populated by all types of devices and people acting as metaphors for different cell functions and enzymes. All professors involved thought it was an excellent idea to approach the whole issue in a brand new and creative way.

Through this treatment this film will be especially useful in schools, who are confronted more and more with children suffering these diseases. Because of the advances in science and diets they now stay alive and have a chance to lead somewhat normal lives. These children require special attention and through this film the viewer should gain an understanding in the lives of these children and the people around them.

More and more disorders are found to have a metabolic error at its core. For this steadily growing group of patients the need for information is increasing, in which this film tries to comply.

The film will be built up as an "exciting, tense" story. That and employing contemporary elements such as THE CELL and the MANGA cartoons will ensure a long shelf life, like its predecessor "A small kiss with big consequences" (1995) which was in use for almost a decade.

#### **ABOUT BOKS vzw (Opdrachtgevers)**



**BOKS vzw** is the main umbrella organization Belgian association of patients suffering from these disorders. Founded in 1995 it has had a remarkable impact in the medical world in Belgium. They were responsible for creating the CEMA's (Centre for Inborn Metabolic

Disorders) which are unique in the world.

#### **ABOUT THE MAKER...**

This will be the second film on this subject by Istvan Leel-Össy. Partly because of his own experience he developed a special sensitivity toward complex subjects such as these films. He makes a point of making clear to the viewer what seems complex and intricate. Because of the creative input and slightly unconventional approach the previous film has proven useful, even after 9 years! Only because of the many new developments the need arose for a new production, reflecting the present situation.

#### **Participating medical professionals:**

- ?? Prof Dr JAEKEN; KU Leuven
- ?? Prof CARTON; UZ Gent
- ?? Prof Dr De MEIRLEIR ; UZ Jette
- ?? Prof CASSIMAN ; KU Leuven
- ?? Prof. Dr. Eijskens ; Paola Kinderziekenhuis Antwerpen
- ?? Prof. CORBEEL; ZH Jan Portaal Vilvoorde
- ?? Dr. And van der PLOEG; Sofia Childrens Hospital Rotterdam (NI)

#### **Industry and pharma:**

- ?? Actelion (Pharma)
- ?? Genzyme (Pharma)
- ?? TKT5s (Pharma)
- ?? Nutricia/Milupa (mainsponsors)