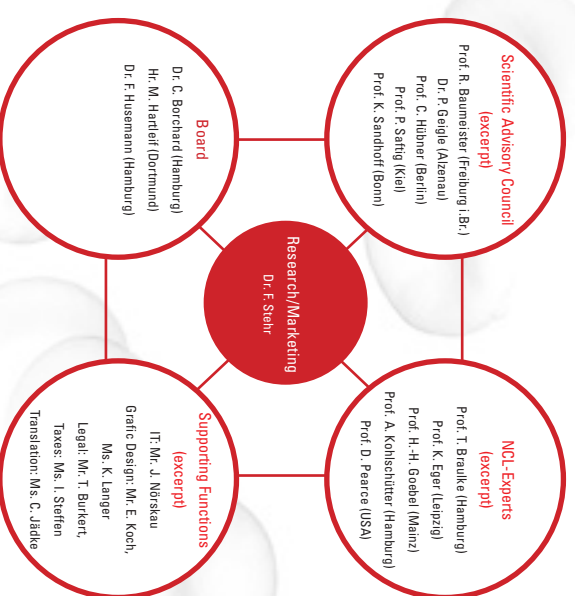


## THE NCL-FOUNDATION

Since its setting-up in 2002 the NCL-Foundation aims at combating rare diseases. Its main emphasis, however, lies on the children's disease NCL (Neuronal Ceroid Lipofuscinosis). When Tim Husemann, the son of the founder, was diagnosed with NCL his father decided to start the foundation.

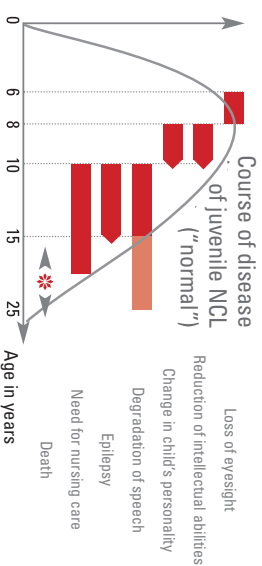
In order to clarify the mechanism of the disease and to develop possible cures, a scientific NCL-network is being established and promising research projects are being initiated. The networking between researchers of different disciplines is supported by NCL-congresses organized by the NCL-Foundation. The third NCL-Congress took place for the first time with international attendance (GB, SF, USA) in order to bundle up the worldwide knowledge about NCL.



The first Ph.D. scholarship to be financed by the NCL-Foundation at the University Hospital Hamburg-Eppendorf is meant to give an impetus to privately financed research concerning NCL. Furthermore, research applications reviewed by the Scientific Advisory Council are passed on to cooperating foundations to cover the missing fields of research as fast as possible. It is a **"National Contest for Life"**.

## WHAT IS NCL?

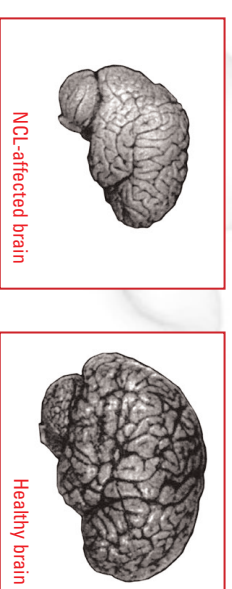
Neuronal Ceroid Lipofuscinosis (NCL) is the most common neurodegenerative disease in childhood. This fatal metabolic disease causes nerve cells to gradually die.



Schematic diagram of the average course of jNCL

The NCL-Foundation concentrates on juvenile NCL. Here the initial symptoms appear between 4 and 10 years of age. The first symptom is rapid loss of eyesight. There then follow various degenerative changes in intellect and speech. Epileptic seizures lead to motoric disabilities. When the disease reaches an advanced state NCL-children are confined to bed. After years of suffering at about 20-30 years of age death is inevitable.

The disease is based on a genetic defect, which is recessively inherited. The affected gene CLN3 lies on the 16th chromosome. The defect leads to an accumulation of insoluble clots within the cell. The cell cannot be cleaned from endogenous toxic substances from the body's daily energy production; it gets overburdened and dies. This concerns mostly the nerve cells of the brain.



The volume of the brain of a patient suffering from juvenile NCL is considerably smaller than that of a healthy individual because of the pathological dying of the nerve cells.

## NCL-RESEARCH

Medical science still needs to understand the NCL disease to finally be able to cure it. In order to fathom all facets of this disease the composition of the accumulated clots needs to be completely analyzed to identify the affected metabolic pathway. Once these results are available the type of protein and the manner of administration can be decided on. A causal therapy including gene correction and / or gene therapy might be possible. These strategies, however, are still under trial or have not been tested with regard to NCL.

Basic research can simulate the course of the disease with different modeling systems. This then can provide the answers to find new active substances that can inhibit the continuous destruction of the delicate nerve cells. Medication for Alzheimer's or Parkinson's that has already been approved could be promising in this context. They still have to be tested for NCL. The NCL-Foundation paves the way to do so.

## NCL Areas of Research (simplified presentation)



## INFORMATION AND SPONSORING

Information and public relations are main objectives of the work of the foundation. Only public awareness allows for long-term prevention! Different events were organized in 2004 to support the foundation.



On 8 May 2004 the "Leo-Service-Day" took place for the benefit of the NCL-Foundation. Around 200 Leo-clubs organized different donation events all over Germany. Thanks to the Lions Club-Zweibrücken this annual report was made possible.

In the middle of the year over 50 hairdressing salons in Hamburg collected donations for the NCL-Foundation during the campaign "Haircut with Heart". The Hamburg Round Tables helped put into action this campaign.



"Cooking for Kids": This was the motto of the first charity dinner organized by the NCL-Foundation. Celebrities known from TV, as Lou Richter (Sat.1) and Dr. Martin Wilhelm (NDR) actively sponsored together with 40 guests the work of the foundation. TV chef Tim Mälzer (VOX) provided for culinary delights. The evening was under the patronage of Dr. Jörg Dräger, Senator for Health and Research in Hamburg.

### Media

In 2004 the foundation was again present on TV and in newspaper articles. Aside from newspaper articles (Hamburger Abendblatt, Wams) and magazine articles, NCL was also featured in TV shows - "Hallo Deutschland" ZDF, "Taff" Pro7.

### Donators and Sponsors

Many private donators have continued to support the work of the foundation. Some have used private events (weddings, christenings, etc.) to ask their guests for donations instead of gifts. We also like to thank the new business sponsors who support children suffering from NCL with single donations. Amongst these companies are: Accenture, Die Bahn, E-Plus, HanseNet, Management Circle, Microsoft.

Special thanks to all donors, sponsors and comrades-in-arms for their support!

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## NCL-ACCOUNT FOR DONATIONS

Account number: 1059 / 22 30 30

Bank code: 200 505 50

Bank: Hamburger Sparkasse

Purpose: Name and address of the donor; donor reference if required.

For all donations exceeding 100 you will receive a receipt for tax purposes. We therefore ask you to state your address in the purpose field.



## THE NCL-FOUNDATION ANNUAL REPORT 2004

Since its setting-up in 2002 the NCL-Foundation is combating the deadly disease Neuronal Ceroid Lipofuscinosis. Several important milestones were reached in 2004 concerning both public relations and research:

- Third German NCL-Congress (June 2004) was held with international attendance in Kronberg/Ts.
- Co-financing of an NCL Ph.D. scholarship at the University Hospital Hamburg-Eppendorf (UKE)
- Integrating "NCL" as part of the international EU research project "APOPIS" (2004-2006)
- Expansion of the NCL-network by integrating researchers from the first NCL project all over Europe "NCL models" (2004-2006)
- Different campaigns for public awareness, such as Leo-Service-Day, "Haircut with Heart", Charity Dinner "Cooking for Kids"

Despite these advancements there is still no cure for NCL. That is why we continue the battle against this deadly children's disease.



2004