

# **Neuroborreliosis in Childhood**

## **- Facts and Fictions**

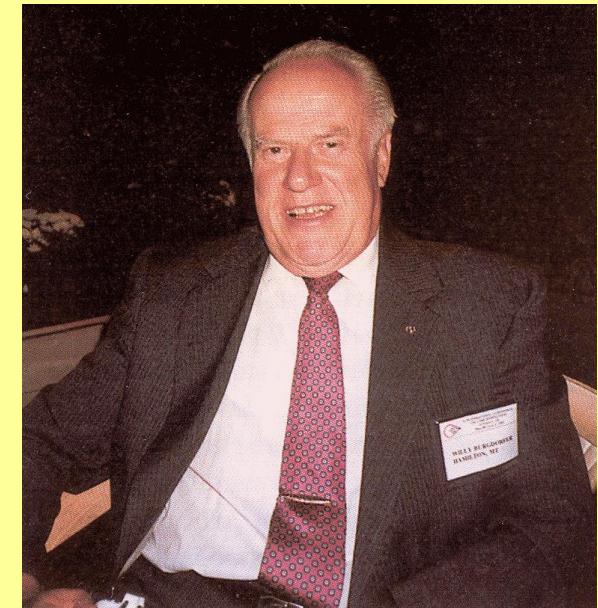
Hans-Jürgen Christen  
Kinderkrankenhaus auf der Bult/Hannover



6th Danish Pediatric Infectious Diseases Symposium  
Korsør, October 5-6th, 2012



**Willy Burgdorfer,  
1982**



**Lyme disease**  
**- discovered in children  
suffering from arthritis  
(Allan Steere 1976)**

**35 years of Lyme disease:**

**What are the facts in childhood?**

## **Fact 1:**

**The risk of becoming sick  
with Lyme disease is rather low.**

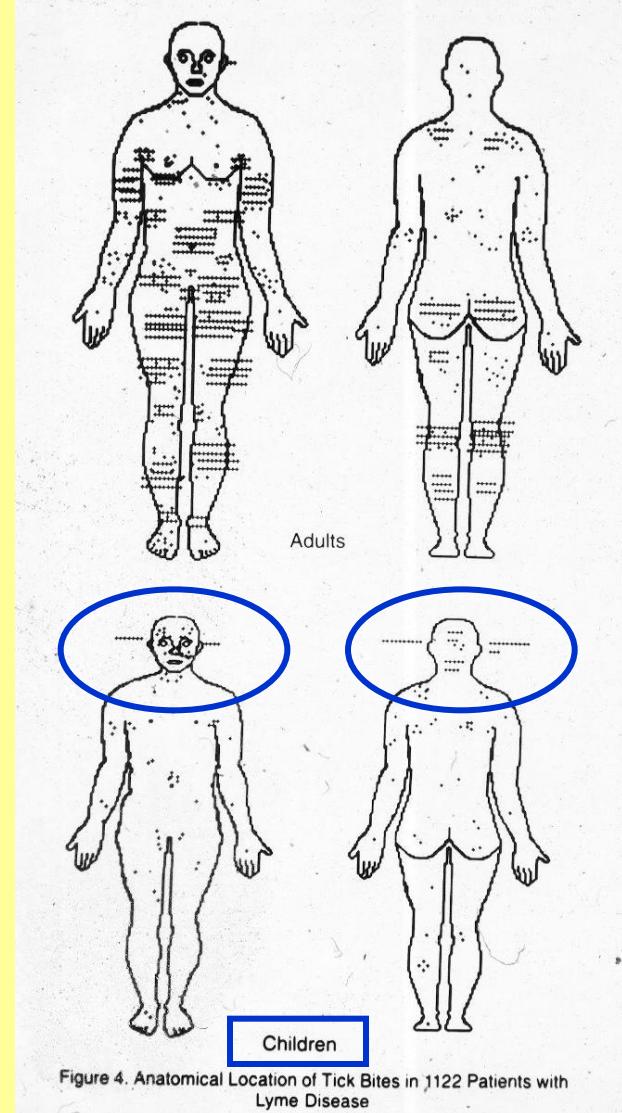
## The risk of becoming ill after a tick bite in an endemic area:

- Tick bites **very frequent**
  - Infected ticks **frequent (30%)**
  - Infection rate **low (1,5-6%)**
  - Illness **very rare (0,3-1,5%)**
- 

## Fact 2:

In children

- in contrast to adults -  
the head-neck region is  
mainly affected by  
tick bites.



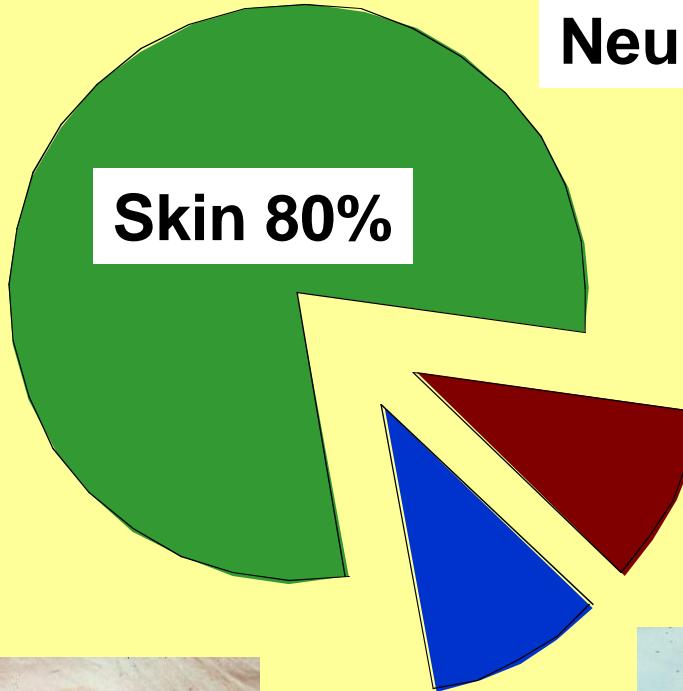
## Fact 3:

**Lyme disease as a multisystem disorder  
is a chameleon with a variable phenotype  
in children – but ...**

## Fact 4:

**Lyme disease as a multisystem disorder  
is a chameleon with a variable phenotype  
in children – but mainly it is a skin disorder.**

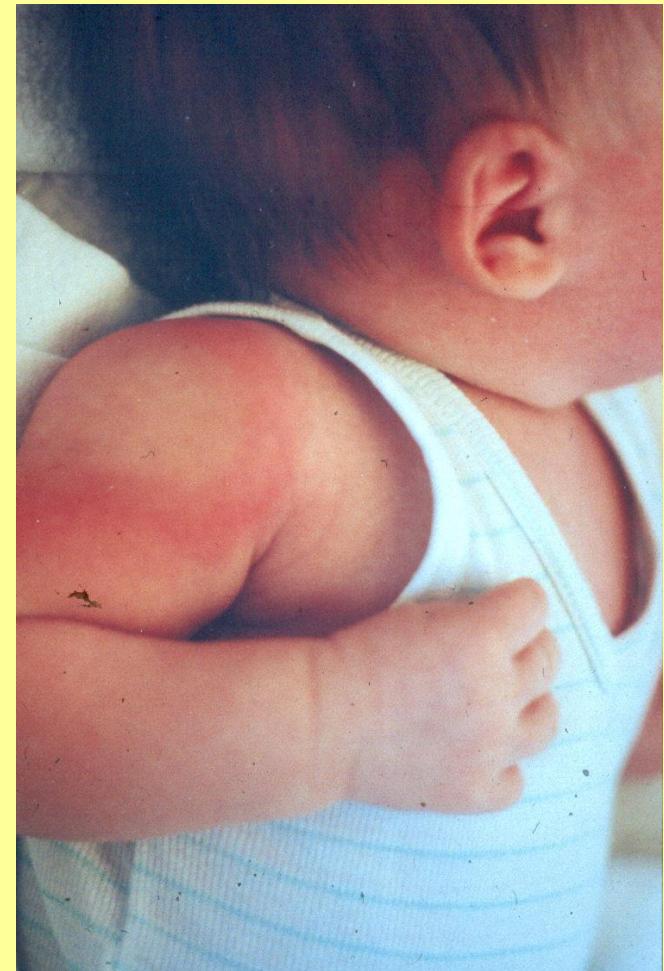
# Lyme-Borreliose



# Erythema migrans (homogeneous form)



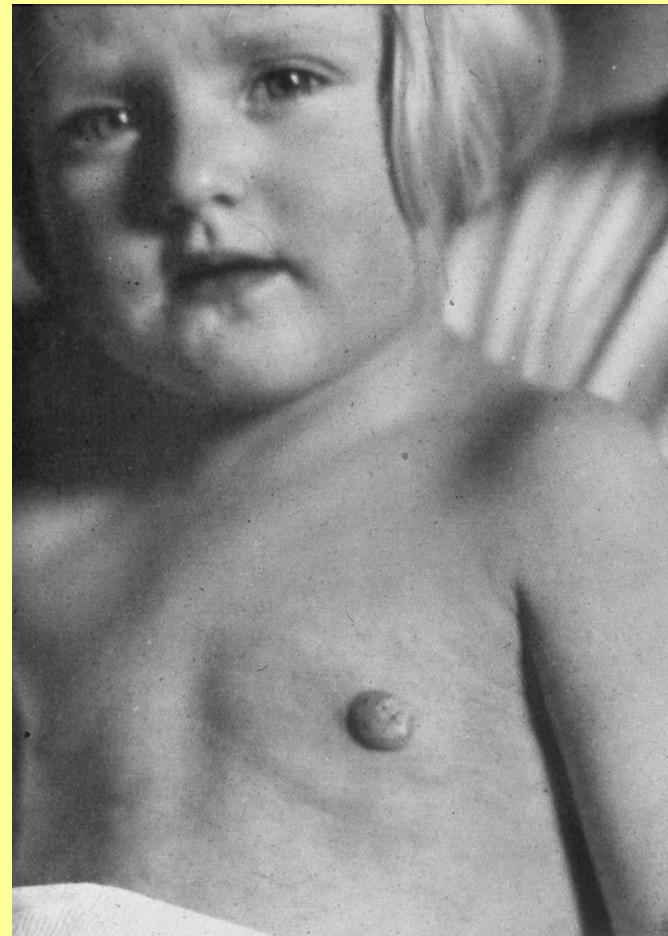
# Erythema migrans (ring-shaped form)



# Erythema migrans (multiple lesions)

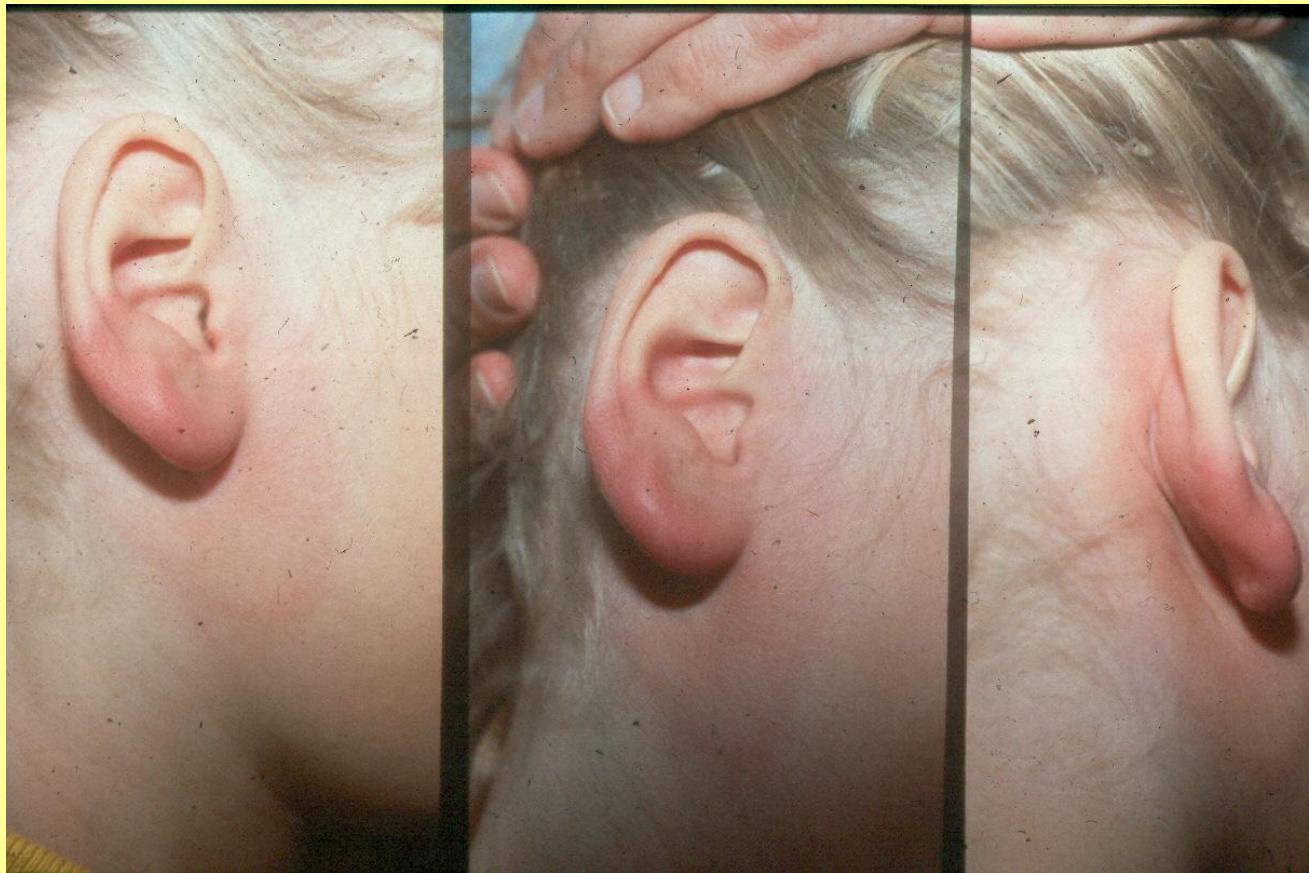


# Borrelial lymphocytoma



Lymphadenosis benigna cutis mamillae  
et areolae mammae (BÅFVERSTEDT, 1943)

# Borrelial lymphocytoma with associated erythema migrans

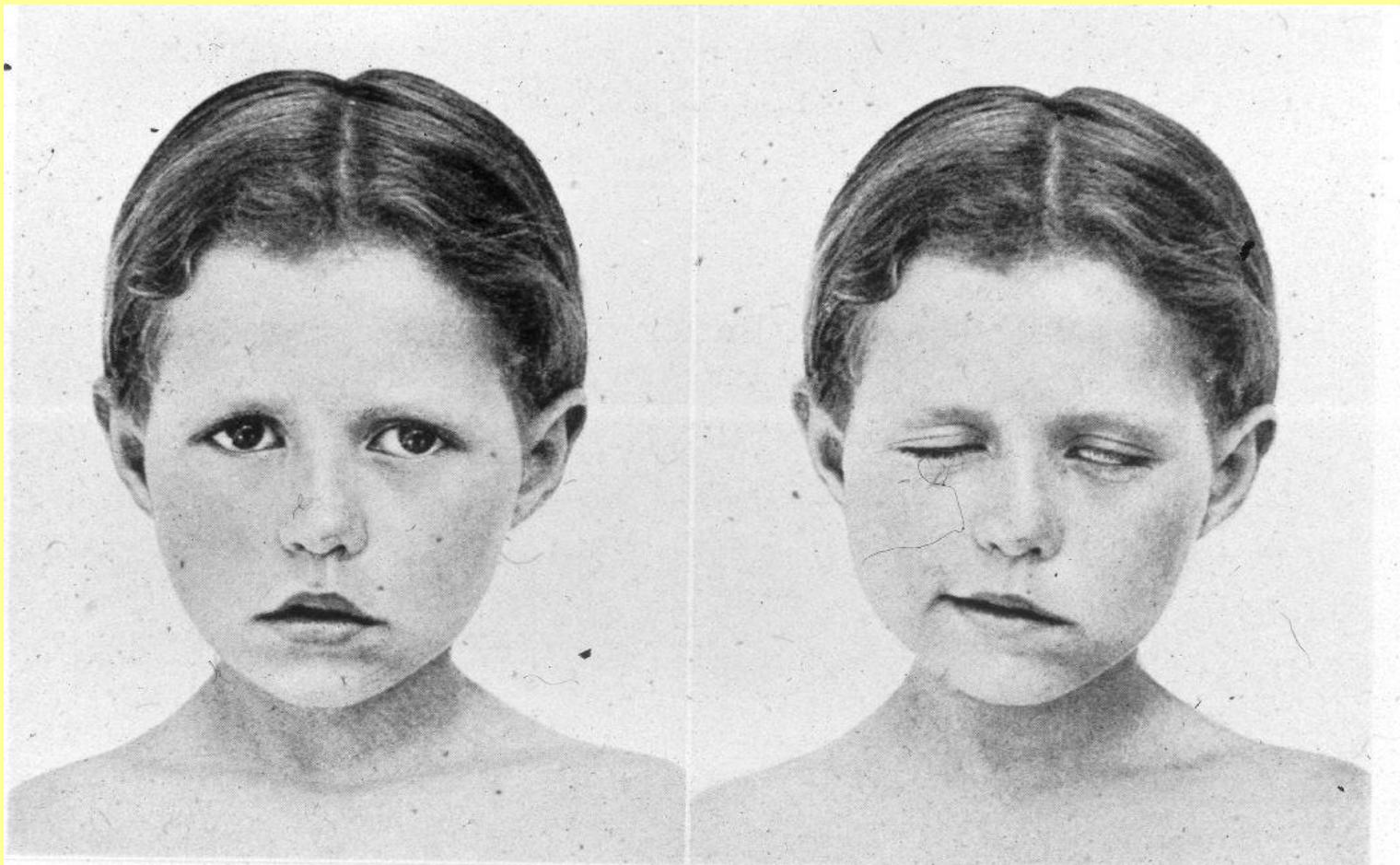


# **Focus on neuroborreliosis**

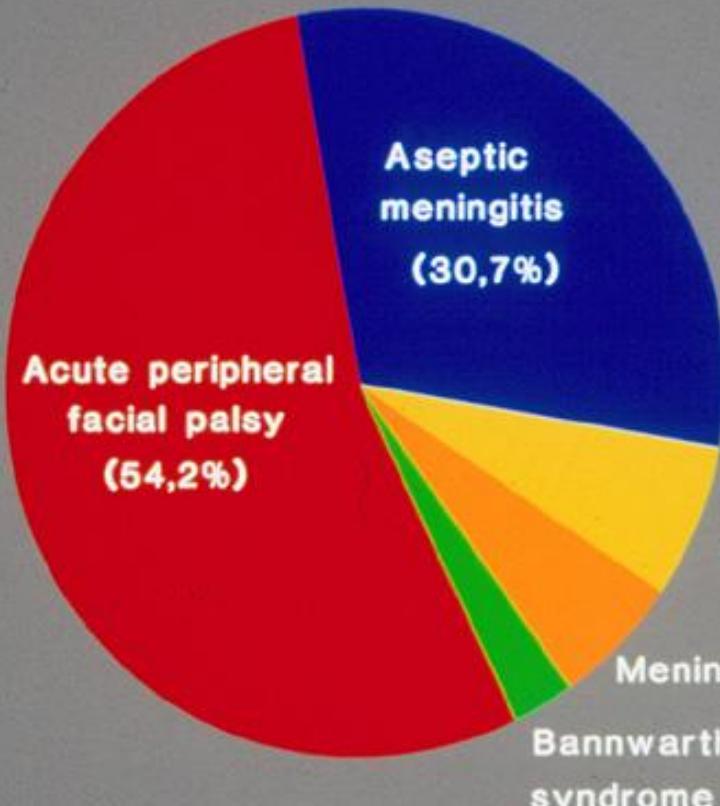
## Fact 5:

**Neuroborreliosis is an old disease  
which was re-discovered 35 years ago.**

# Lyme-Borreliose



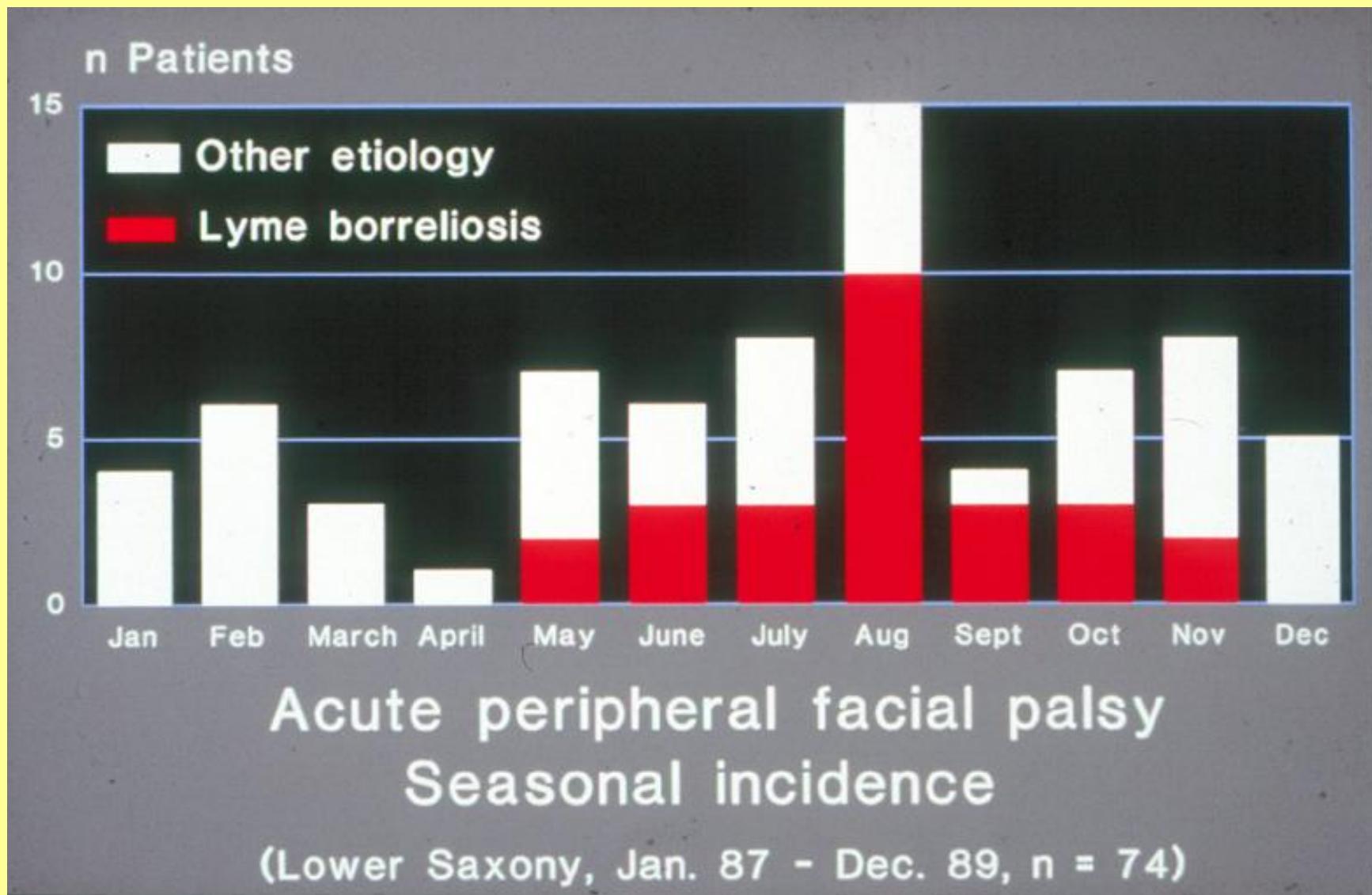
Fazialisparese bei allergischer Meningitis (8 J.)  
(PETTE, 1942)



(n = 179)

Other syndromes : Abducens nerve palsy  
: Oculomotor palsy  
: Acute ataxia  
: Guillain-Barré-syndrome  
: Headache  
: Pseudotumor cerebri

## Neuroborreliosis in childhood - Clinical spectrum (Diagnostic criteria: IgM antibodies in CSF)



## Fact 6:

**Lyme disease is mainly a clinical diagnosis  
rather than a laboratory one.**

# 4 diagnostic columns:

## 1. History, but ...

**... the tick bite is an uncertain criteria,  
it means neither a proof  
nor an exclusion of Lyme disease.**

# 4 diagnostic columns:

- 1. History**
- 2. Clinical signs (→ Skin)**

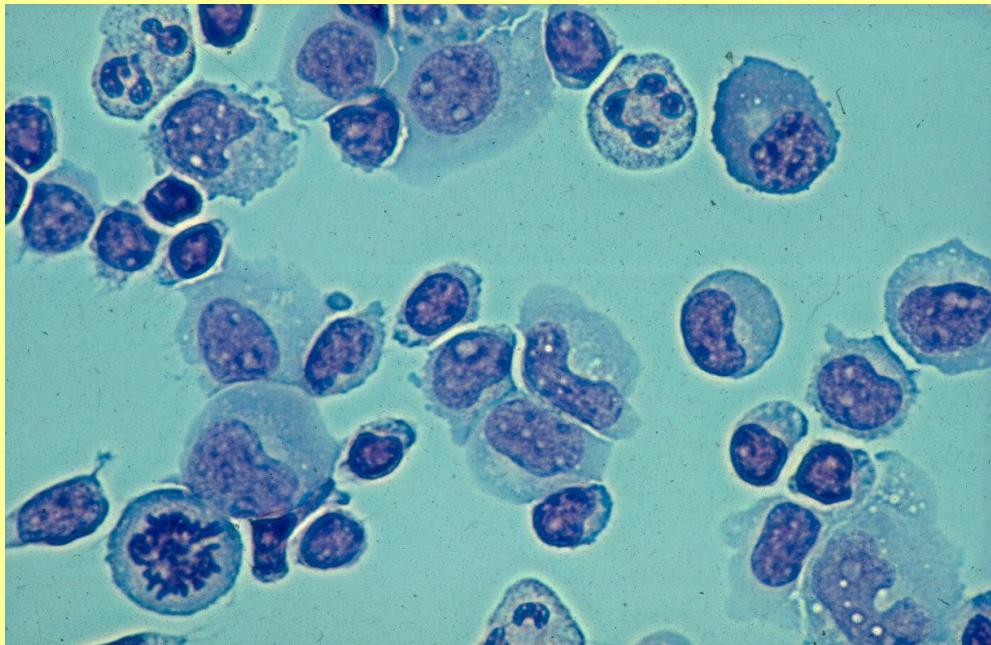
# 4 diagnostic columns:

- 1. History**
- 2. Clinical signs (→ Skin)**
- 3. CSF findings (→ neuroborreliosis)**

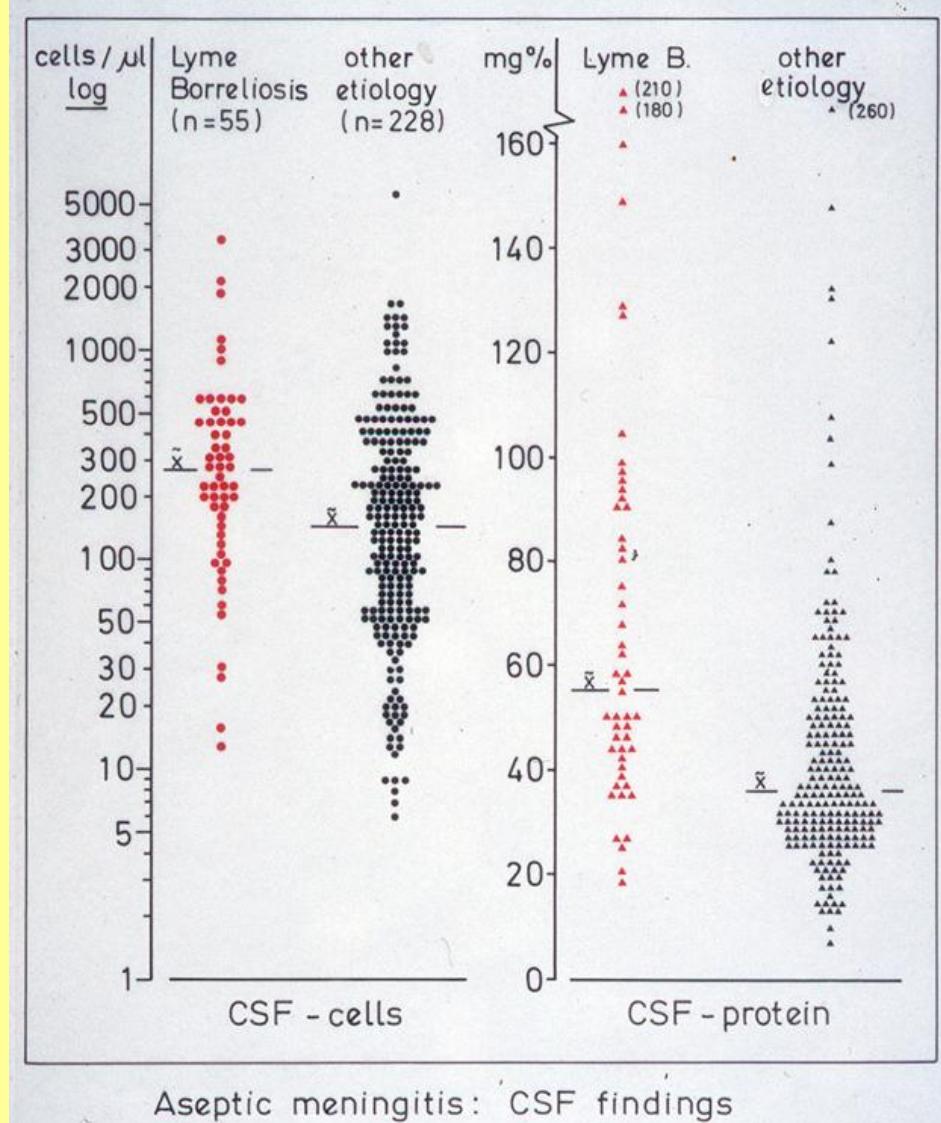
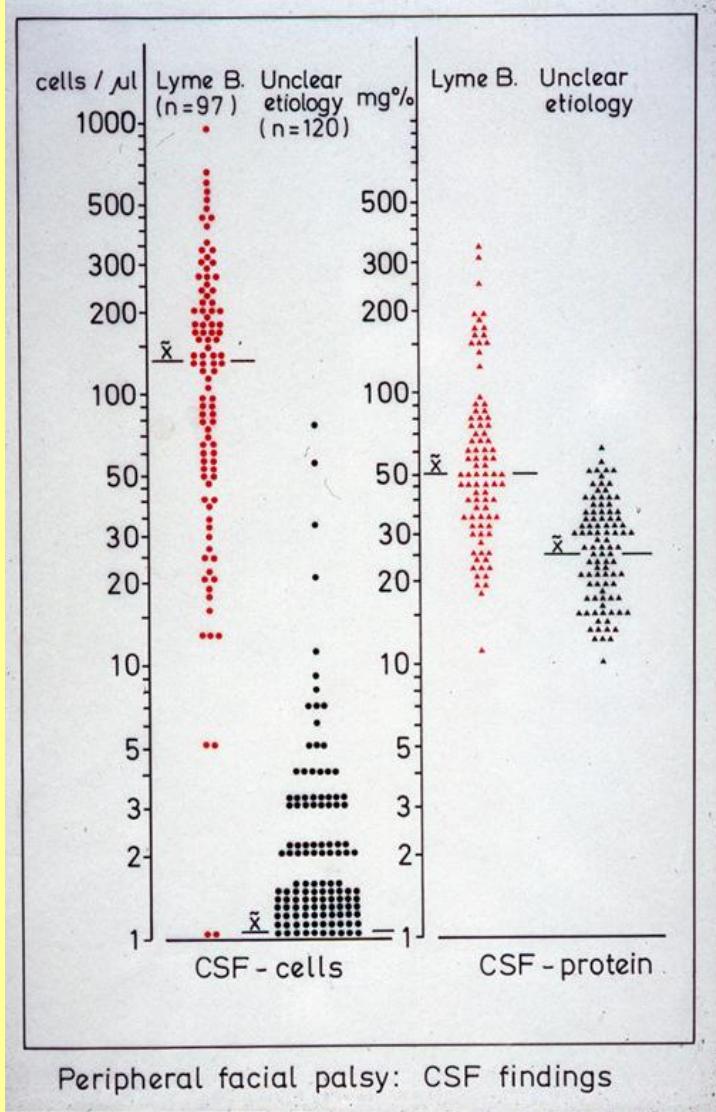
# **Neuroborreliosis:**

## **Lymphocytic CSF pleocytosis**

### **,conditio sine qua non ‘**



# Lyme Borreliosis: CSF findings in neuroborreliosis



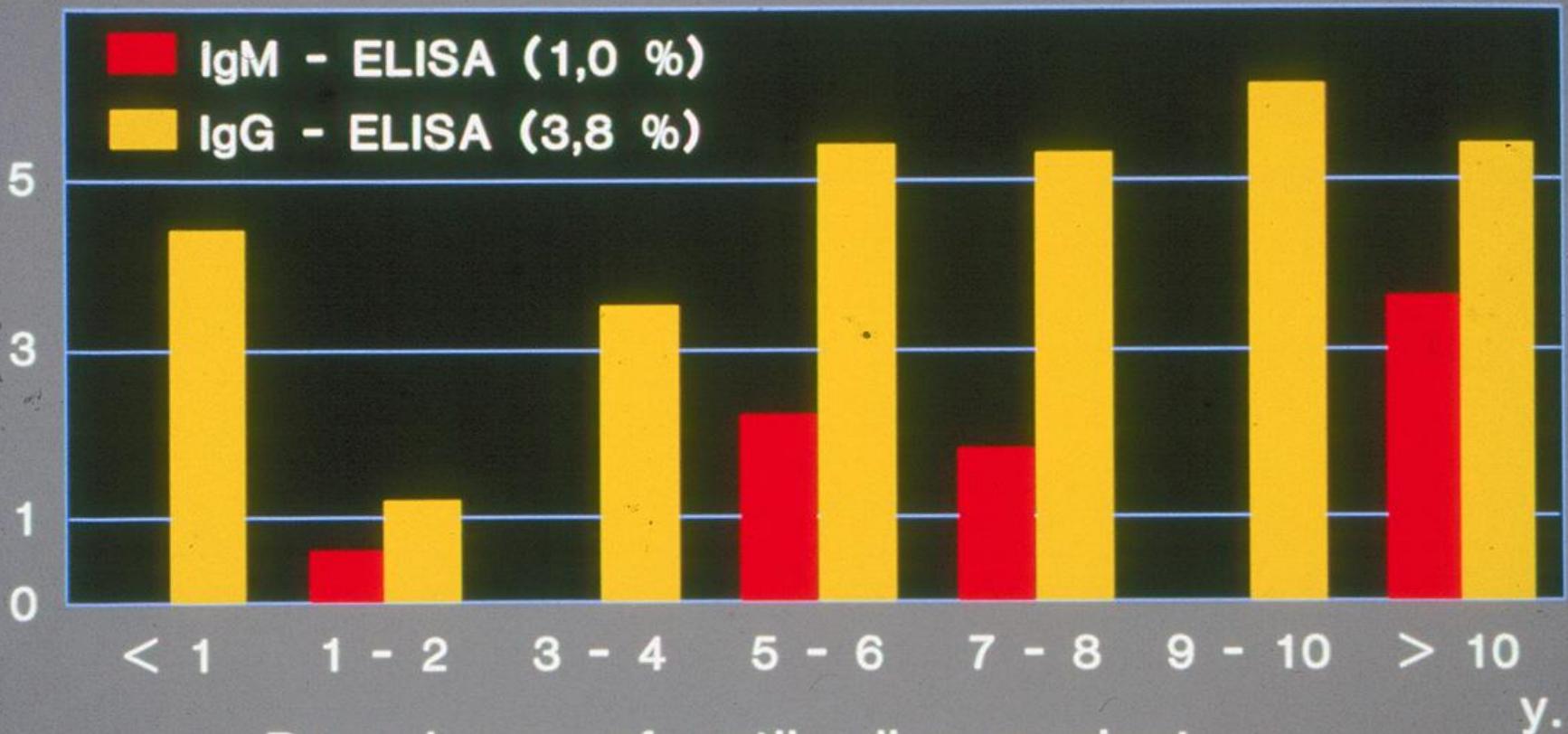
# 4 diagnostic columns:

- 1. History**
- 2. Clinical signs (→ Skin)**
- 3. CSF findings (→ Neuroborreliosis)**
- 4. Antibody testing**

## Fact 7:

**Positive antibody results  
do not proof Lyme disease  
nor do negative results exclude it.**

% of age group



Prevalence of antibodies against  
Borrelia burgdorferi in childhood  
(Lower Saxony, Nov. 1987 - Oct. 1989, n=580)

## **Two clinically important questions:**

- 1. Headaches and neuroborreliosis:  
How to proceed?**
  
- 2. When can neuroborreliosis be  
regarded as excluded?**

# **Subacute Borrelial meningitis**

=

**Headaches plus ...**

## Presenting symptoms: Headaches + ...

- ill since 5 days
- headaches, vomiting,  
abdominal pain
- no meningism, but ...

... marked stiffness of the spine  
(distance to the floor 30 cm)

## Presenting symptoms: Headaches + stiffness of the spine

**CSF**      **cells 826/ $\mu$ l, protein 115 mg%,  
glucose-ratio 0,36 (!!!)**

### Antibody results:

**Serum**      **IgM ELISA -**  
**IgG ELISA -**

**CSF**      **IgM ELISA -**  
**IgG ELISA (+)**

## Course: immediate remission with antibiotic therapy

## Neuroborreliosis: Subacute meningitis – Signs and symptoms

	head- aches	tired- ness	inappe- tence	arthralgia	meningism
Carolin	+			+	-
Sven	+	+			-
Moritz	+	+			-
Sahra	+		+		-
Christian	+		+		-

## Neuroborreliosis: Subacute meningitis - CSF findings

	leukocytes ( $\mu$ l)	protein (mg/dl)	Glucose CSF/blood	pressure (cm H <sub>2</sub> O)
Sahra	15	normal (24)	0,49	42
Carolin	112	95	0,58	-
Sven	241	56	-	-
Moritz	225	74	0,56	-
Christian	280	97	-	-

## Neuroborreliosis: Subacute meningitis – antibody results

	Serum		CSF	
	IgM	IgG	IgM	IgG
Sahra	+	+	(+)	(+)
Carolin	+	+	(+)	+
Sven	+	(+)	(+)	(+)
Moritz	+	-	+	(+)
Christian	+	+	(+)	(+)

### Diagnostic clues in neuroborreliosis:

1. **Nightly headaches**
2. **Occipital headaches**
3. **Inappetence**  
("anorexia nervosa-like syndrome", Pachner et al. 1986)
4. **Stiffness of the spine (?anteflexion of the trunk)**
5. **Papilledema**
6. **CSF opening pressure ↑**
7. **Bilateral facial palsy (highly suspicious)**

## **First question:**      **Headaches and neuroborreliosis - How to proceed?**

- 1. Focused history taking**
- 2. Focused clinical examination**
- 3. Antibody results in the serum  
= useful screening parameter**
- 4. If antibody testing in serum is positive  
CSF examination („gold standard“)**

## **Second Question:**

**When can neuroborreliosis be regarded as excluded?**

### **Problem:**

**Suggestive neurological signs and symptoms, but normal CSF findings**

## Conclusions:

1. ... to early isn't good either!

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- 1. ... to early isn 't good either!**
- 2. Serial antibody testing sometimes is essential  
to demonstrate seroconversion  
(Albisetti et al. 1997)**

## Second Question:

**When can neuroborreliosis be regarded as excluded?**

- 1. No seroconversion after 4 weeks**
  
- 2. Normal CSF findings in a symptomatology which is already persisting for several weeks**

## Fact 8:

**Lyme disease is a well treatable disorder  
with a favourable prognosis.**

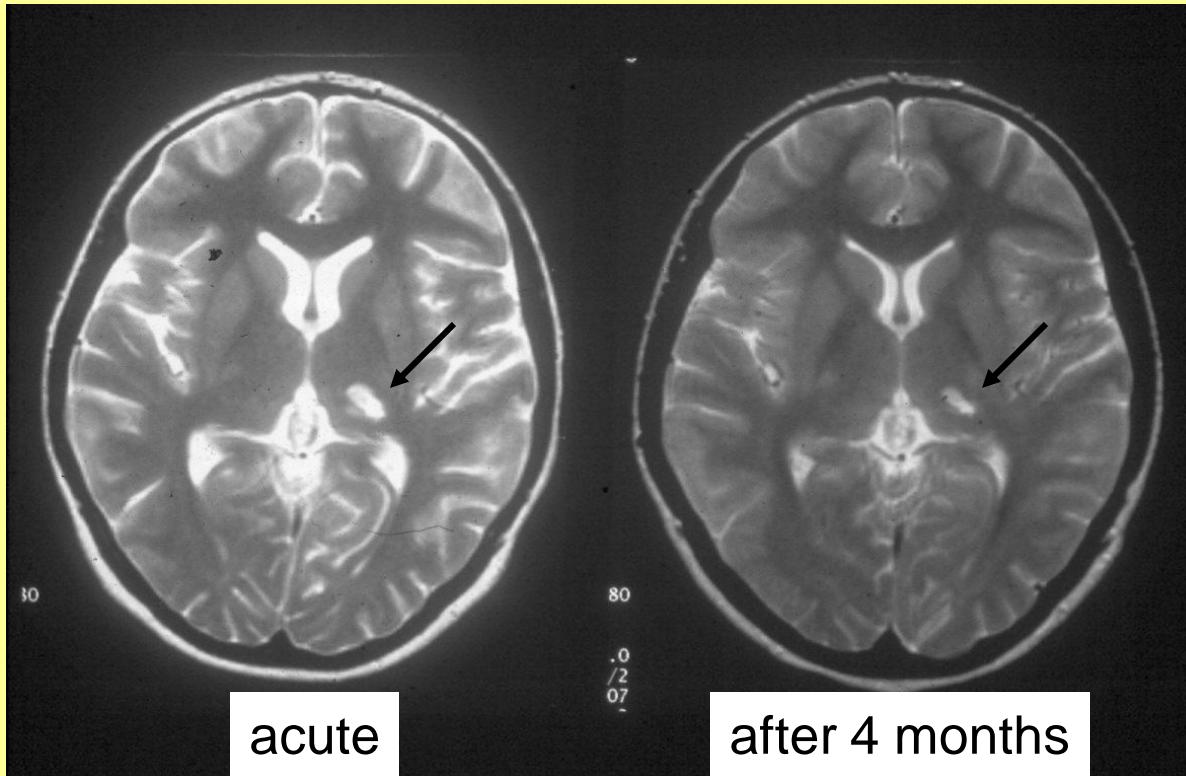
# **Therapy of neuroborreliosis:**

**Doxycycline serious treatment option  
in early neuroborreliosis in children > 8 y.  
with the advantage :**

- **oral application**
- **low costs**

**Fact 9: Neuroborreliosis is a well treatable disorder,  
but has to be taken seriously!**

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but has to be taken seriously!**



**Neurovascular complication in a case of  
neuroborreliosis with acute hemiplegia**

## Fiction 1:

**„Tick bites are frequently  
causing neuroborreliosis in children.“**

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**„Tick bites are frequently  
causing neuroborreliosis in children.“**

**No, only in 1 out of 1000 cases of tick bites,  
but look carefully for early tick removal!**

## Fiction 2:

**„Lyme disease is a frequent cause of chronic headaches in children.“**

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**„Lyme disease is a frequent cause of chronic headaches in children.“**

**No, but look carefully for additional signs and symptoms!**

## Fiction 3:

**„Antibody testing for Lyme disease is  
the decisive tool for diagnosis or exclusion.“**

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**„Antibody testing for Lyme disease is  
the decisive tool for diagnosis or exclusion.“**

**No, but it may serve as a screening tool  
and support the diagnosis!**

## Fiction 4:

**„Neuroborreliosis is dangerous  
in children.“**

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**„Neuroborreliosis is dangerous  
in children.“**

**No, but it should be treated consequently  
on the basis of a well-defined diagnosis!**

**Lyme disease rather is over-diagnosed  
than under-diagnosed**

**but ...**

**Lyme disease rather is over-diagnosed  
than under-diagnosed**

**but ...**

**... in case of confirmed Lyme disease  
a consequent antibiotic treatment is mandatory.**