Challenges facing different cultures

Anne Malfroot, M.D.,Ph.D.
CF centre UZ Brussel – VUB
Brussels, BE www.mucojette.be

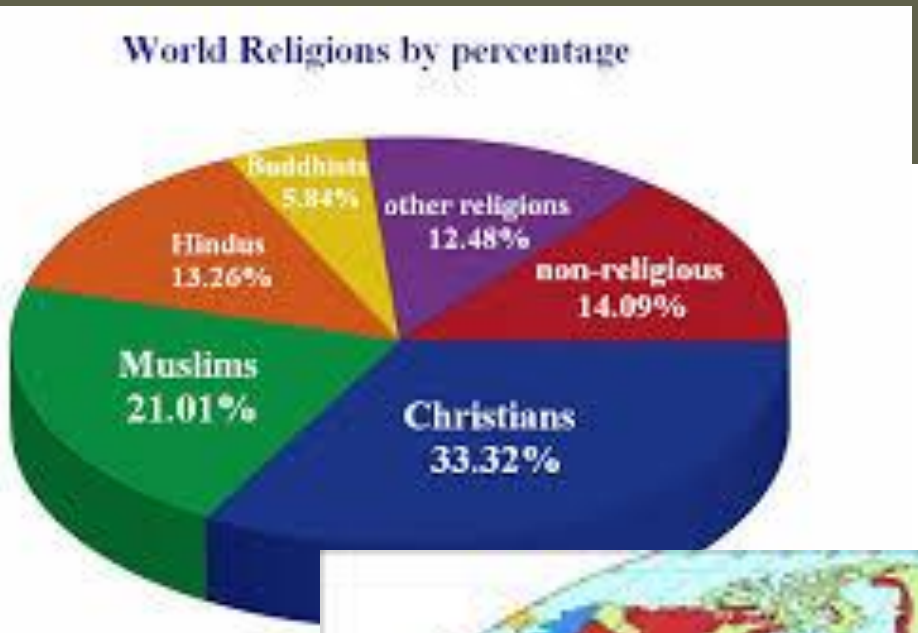
mucojette.be

Vrije Universiteit Brussel

A **religion** is an organized collection of beliefs, cultural systems, and world views that relate humanity to an order of existence. Many religions have narratives, symbols, and sacred histories that aim to explain the meaning of life. From their beliefs about the cosmos and human nature, people may derive morality, ethics, religious laws or a preferred lifestyle.

The practice of a religion may include rituals, sermons, commemoration or veneration (of a deity, gods, or goddesses), sacrifices, festivals, feasts, trances, initiations, funerary services, matrimonial services, meditation, prayer, music, art, dance, public service, or other aspects of human culture.

Religion has also impacted the way in which societies prevent disease and treat people with certain diseases. Examples of society using religious beliefs to prevent disease are seen in African culture. The AIDS epidemic, leprosy, and the persecution of Jews during the Black Death are all examples in which religious beliefs have allowed society to negatively treat groups of people associated or not associated with certain diseases


World Religions by percentage
Buddhists
5.84%
other religions
12.48%
Hindus
13.26%
non-religious
14.09%
Muslims
21.01%
Christians
33.32%

History of CF: first reports....J.Littlewood

- "origin" in Turkey /Iracq 5000 years ago, spread worldwide
- 1595: Professor Pieter Pauw (Leiden) :
  - I conducted an autopsy on an 11-year old girl said to be bewitched. She had had strange symptoms for eight years. Inside the pericardium, the heart was floating in a poisonous liquid, sea green in colour. Death had been caused by the pancreas which was oddly swollen.
- 1729 Schmidt J G. Book of Folk Philosophy.
  - It was stated that an excessively salty taste to the skin meant a child was bewitched, and will die
- 20st century: scientific reports

Fanconi-1936
Zurich1892-1979

D. Andersen 1938
New York 1901-1963

PREVALENCE

PM. Farrell. JCF 2008. Prevalence CF Europe

Population and prevalence of patients with CF in E.U. countries

	Population in 2004 (thousands)	# CF patients	CF prevalence (per 10,000)	Estimated CF incidence	Source(s)
Austria	8,175	686	0.839	1:3500	Ia, [1]
Belgium	10,348	1065	1.03	1:2850	Ib, IIa, [13,14]
Bulgaria	7,518	170	0.226	1:2500	[13]
Cyprus	776	26	0.335	1:7914	[15]
Czech Republic	10,246	570	0.556	1:2833	Ic, IIa, [14,16]
Denmark	5,413	412	0.761	1:4700	IIa, [14,17,18]
Estonia	1,342	83	0.618	1:4500	[17]
Finland	5,215	64	0.123	1:25000	Id, [17,19]
France	60,424	4533	0.750	1:4700	Ie, IIa, IIe, [1]
Germany	82,425	6835[a]	0.829[a]	1:3300	If, IIa, [14,16,20]
Greece	10,648	555	0.521	1:3500	Ig, [14]
Hungary	10,032	410	0.409		Ih
Ireland	3,970	1182	2.98	1:1353	Ii, IIb, [8]
Italy	58,057	5064	0.872	1:4238	IIc, [21]
Latvia	2,306	24	0.104		[7]
Lithuania	3,608	47	0.130		[7]
Luxembourg	463	20	0.431		[7]
Malta	397	23	0.579		IV
Netherlands	16,318	1275	0.781	1:4750	IIa, [22]
Poland	38,580	987	0.256	1:5000	Ij, [1]
Portugal	10,524	285[a]	0.271[a]	1:6000	Ik,[7]
Romania	22,356	238	0.106	1:2056	[23]
Slovakia	5,424	340	0.627	1:1800	IIa, [24]
Slovenia	2,011	66	0.328	1:3000	[7,25]
Spain	40,281	2200[a]	0.546[a]	1:3750	Il, [13,14]
Sweden	8,986	362	0.403	1:5600	IIa, [26]
United Kingdom	60,271	8284	1.37	1:2381	Im, IIa, IId, [9]

The disease was first described in 1936, and its prevalence among Caucasian in Europe and North America is estimated between 1 in1600 to 1 in 2000, it reported to be very rare amongst American Blacks (1 in 17,000) and Orientals (1 in 90,000).

The first Arab child with CF was documented by Salem from Lebanon in 1962. Later cases were reported from Iraq, Kuwait, Palestine, Jordan, Saudi Arabia (report 2003 → 1 in 4,243) and Bahrain (report 1992 → 1 in 7,700).

Exact prevalence in the Arab population is still unknown.

The most common mutation which affects about 70% of Caucasians is DF508 or delta-F508. The 35 mutations tested with the conventional InnoLipa set represent approximately 90% of the mutations found in Caucasian CF mutation carriers.

In other ethnic groups the rate of detected mutations can decrease significantly. For example it is estimated that the mutation detection rate with this method is around 33% for the Turkish population. For the Arabian population only 10 of the recurrent mutations are detectable with the conventional technique

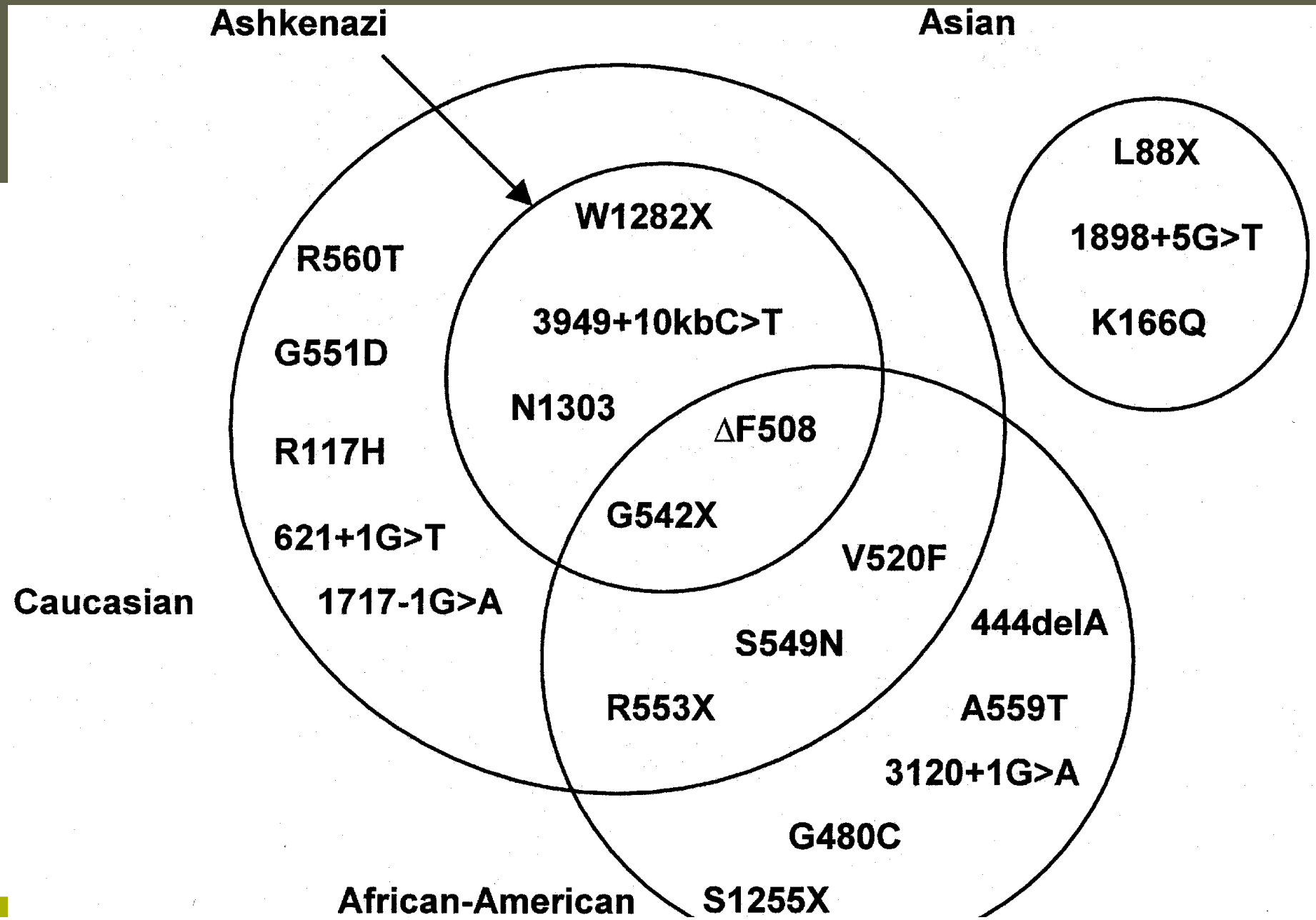


Fig. 1. Distribution of known CFTR mutations amongst different ethnic populations (reproduced with kind permission of Garry Cutting M.D., Johns Hopkins, Baltimore MD, USA).

CFTR mutations in Algeria*, Morocco**

Few data

Lower F508 prevalence

Heterozygotes

Mutations with borderline sweat test Chloride

* JCF (7) 2008: 54-59 / **440-443

Socio-cultural challenges

1. establishing and maintaining patient-professional relationships
2. patient education
3. adherence to treatment
4. care of dying and death

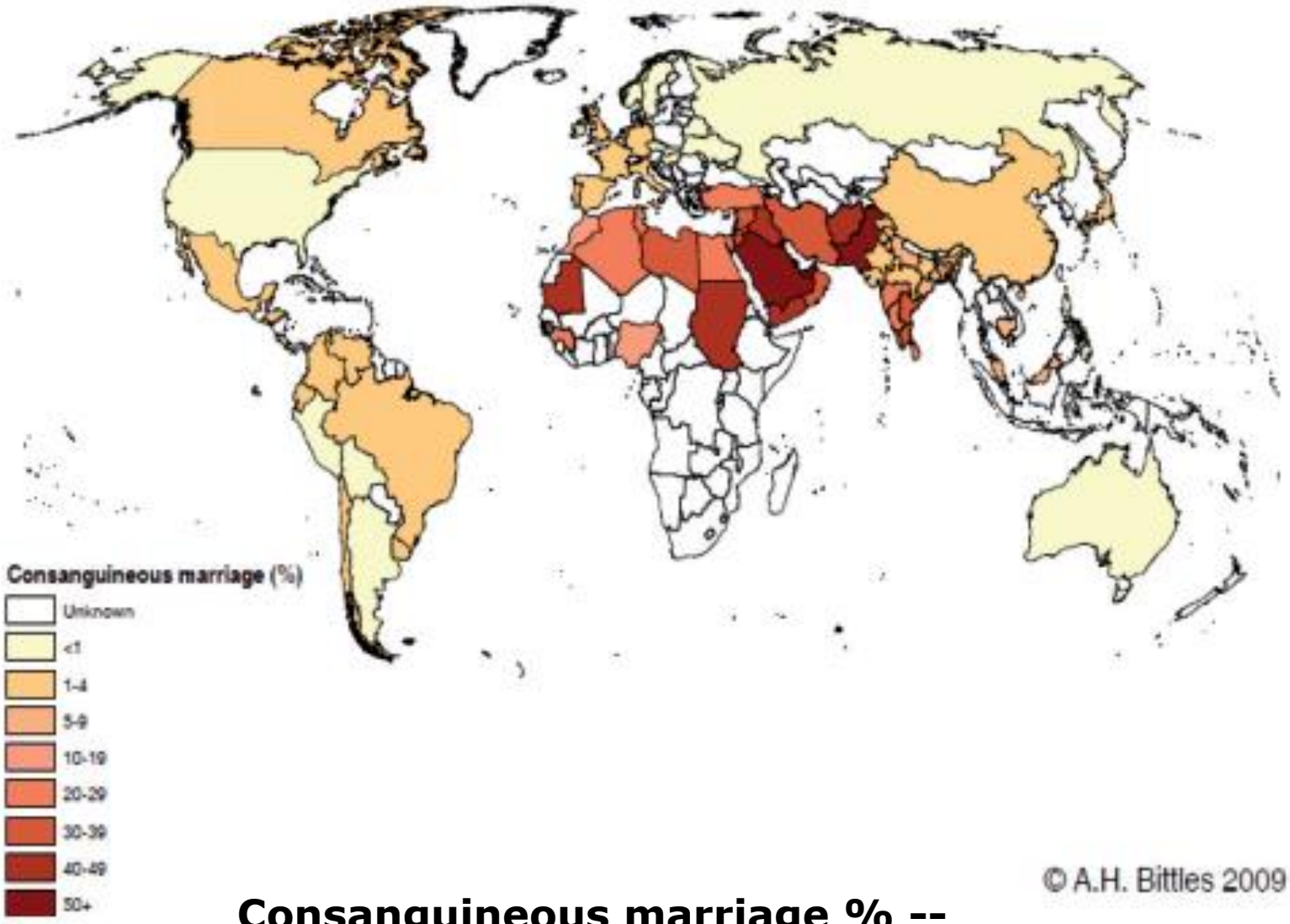
A.J. Duff / Journal of Cystic Fibrosis 2 (2003) 38–41

1. Patient-professional relationship

-communication, delicate issues (consanguineous parents, birth control, prenatal diagnosis, incurable disease…)

-language barrier:

- Avoid help of family members for interpretation
- Try to communicate directly
- Use of professional interpreter



Consanguineous marriage % -- increased risk recessive disorders

2. Patient education

-difficult issues related to CF:

loss of lungfunction despite treatment, incurable disease, infertility, longterm implications

-arranged marriages …

-minimize seriousness of the disease

-unrealistic expectations of adult life with CF

-some aspects of treatment may contravene certain beliefs and values (gastrostomy, portacath, transplantation…)

-encourage contact with patients association

-involving local spiritual leaders

3. Adherence to treatment

-need to spent more time to explain importance of chronic treatment

-dietetic management → religious fasting periods

-pancreatic enzyme replacement by porcine derived enzymes …

-gender separation: male physiotherapist giving treatment to female patients…

-transplantation: position on organ donation

4. Care of dying and death

Specific funeral rituals – open discussion about the family's wishes

-Muslims: when dying --> facing the Qiblah (prayer direction Mecca)

-after death: respect instructions for handling body

Brussels Center

n=180 patients ---(n=95≥18 years)

origin	Morocco	Algeria	Turkey	Pakistan	Egypt
n	8 1family n=3 1family n=2	1	1	1	1

Clinical presentation: mild to severe

Conclusions

Minority groups with different ethnic origins

Immigrants in EU countries (specific per EU country)

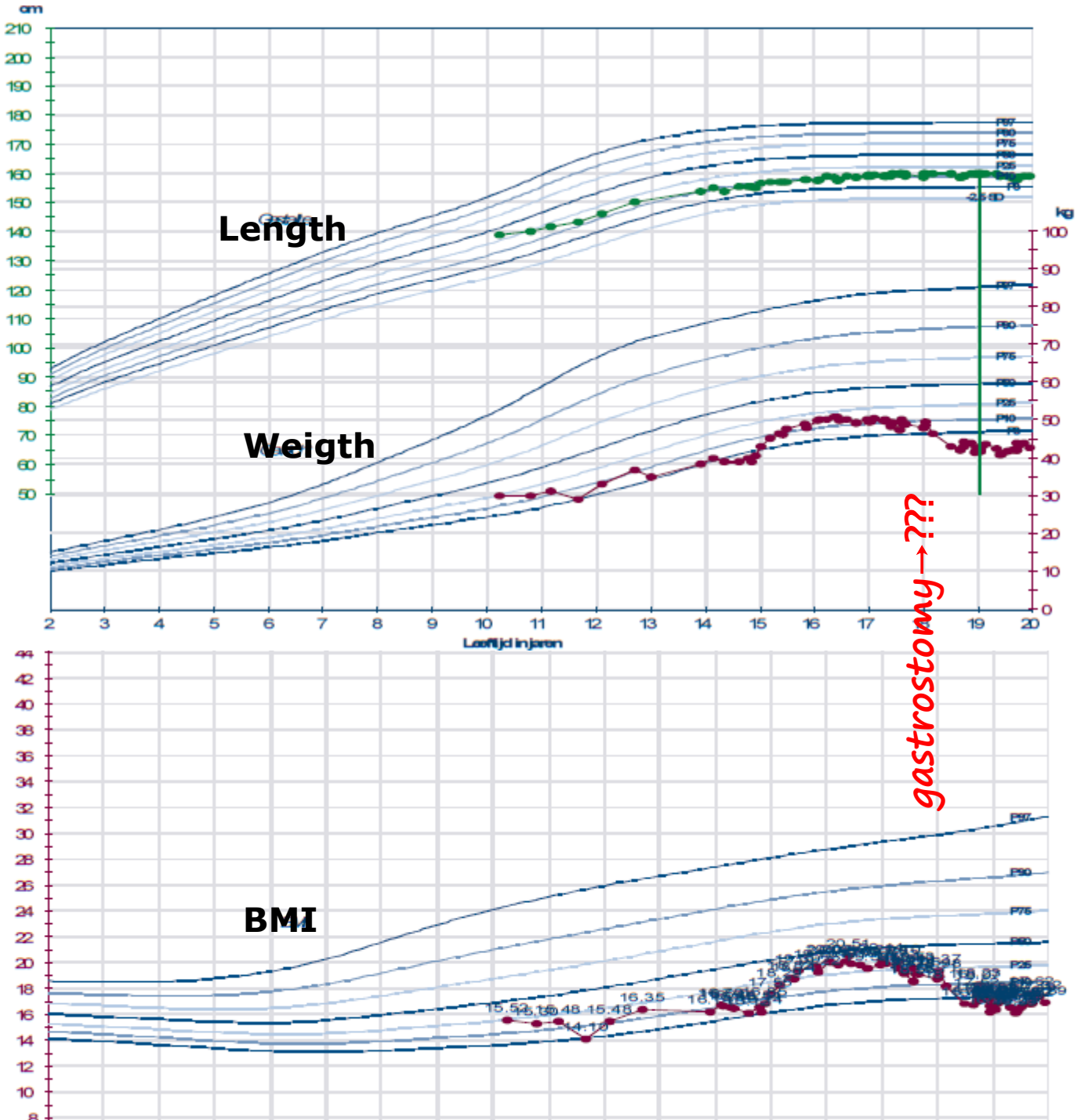
Poorly studied

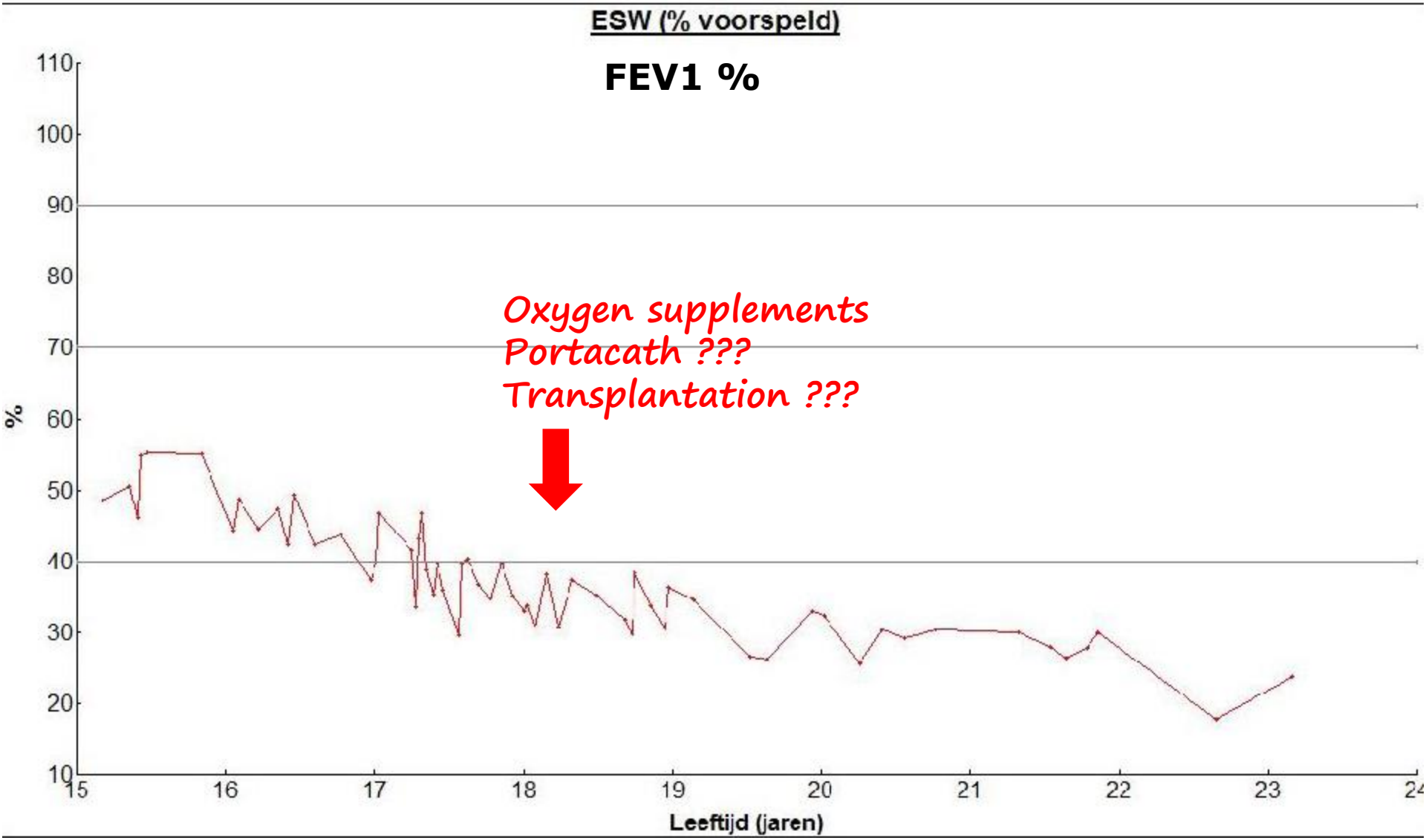
No guidelines for specific care

CASE REPORT

Leila ° 1992

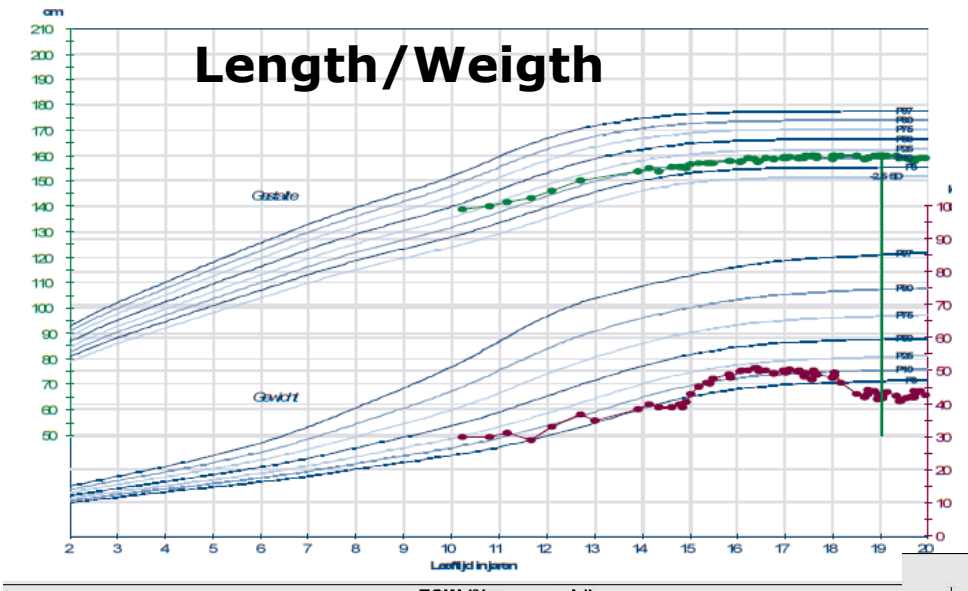
- CF diagnosis by NBS-homozygote F508del
- classic “severe” CF
- diabetes at the age of 15 years
- multiple sclerosis at 16 years
- oxygen supplements at 16 years
- no communication on CF with relatives--> negotiations on treatments, etc…

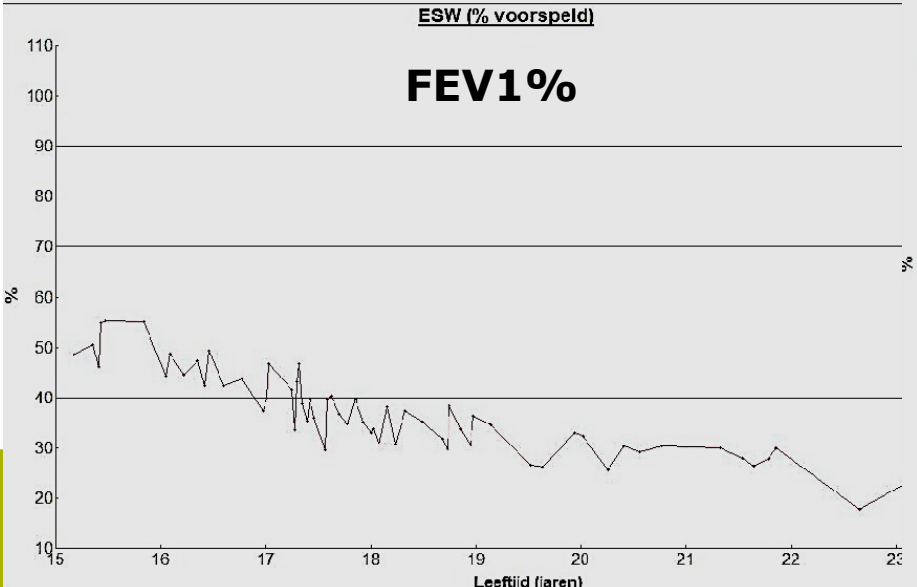

Length
Weigth
BMI
gastrostomy→???
Age in years→

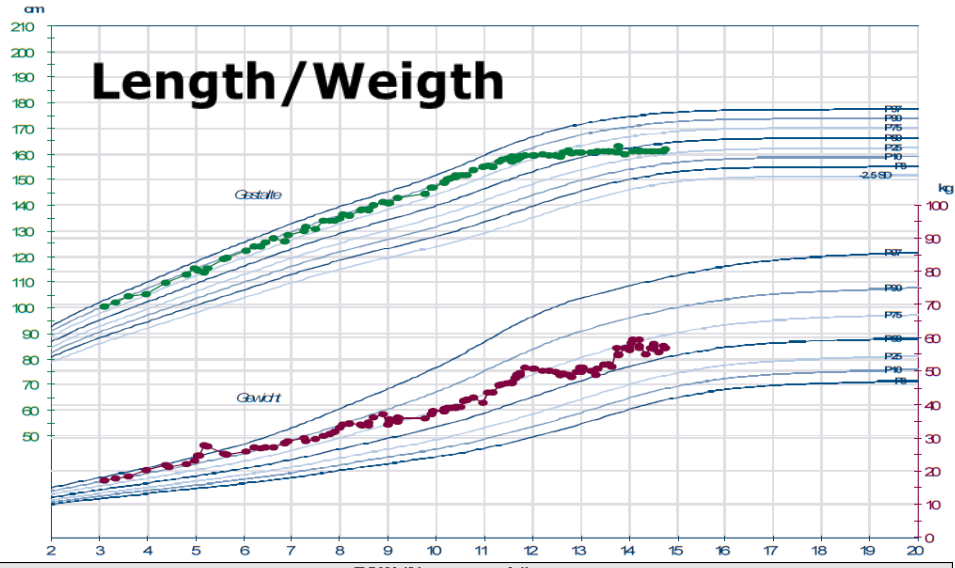

ESW (% voorspeld)
FEV1 %
110
100
90
80
70
60
50
40
30
20
10
%
Oxygen supplements
Portacath ???
Transplantation ???
15
16
17
18
19
20
21
22
23
24
Leeftijd (jaren)

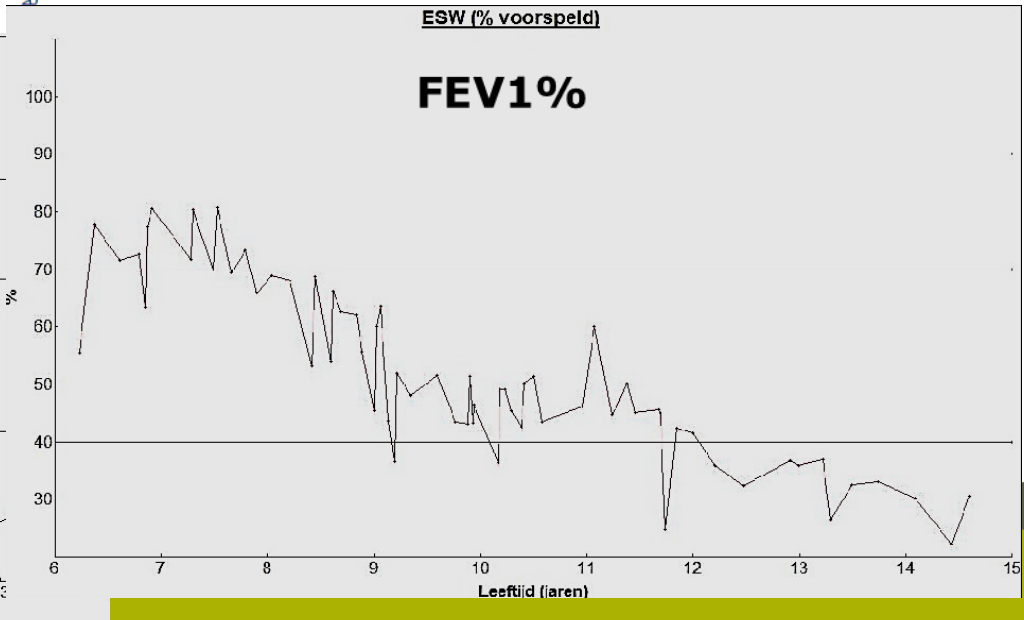
age in years

Leila °1992 Nadia °2000

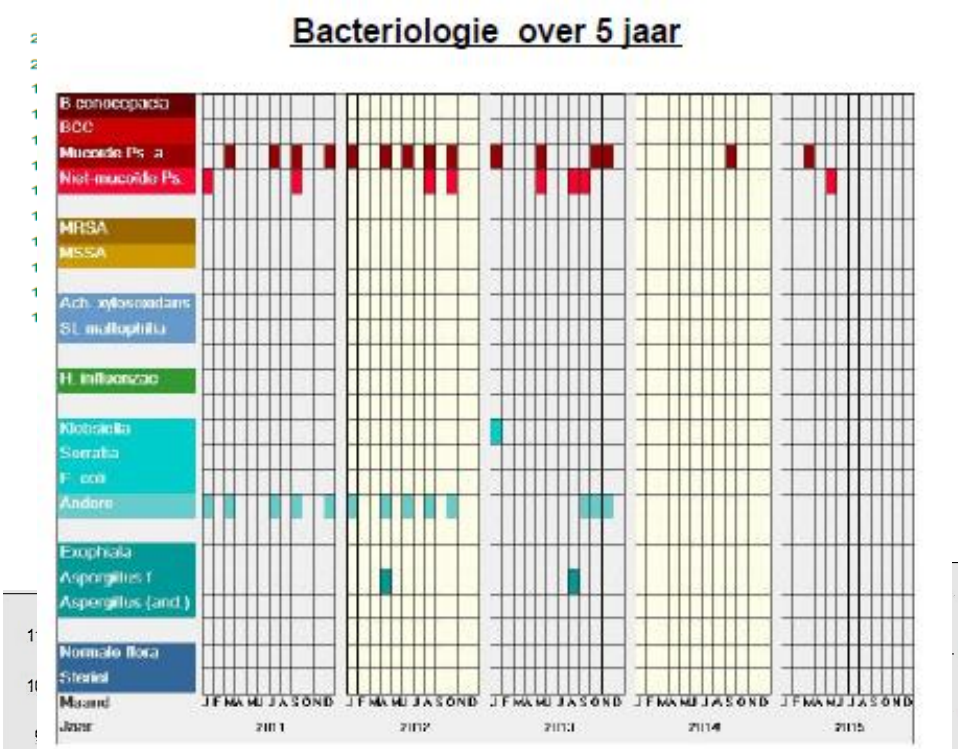


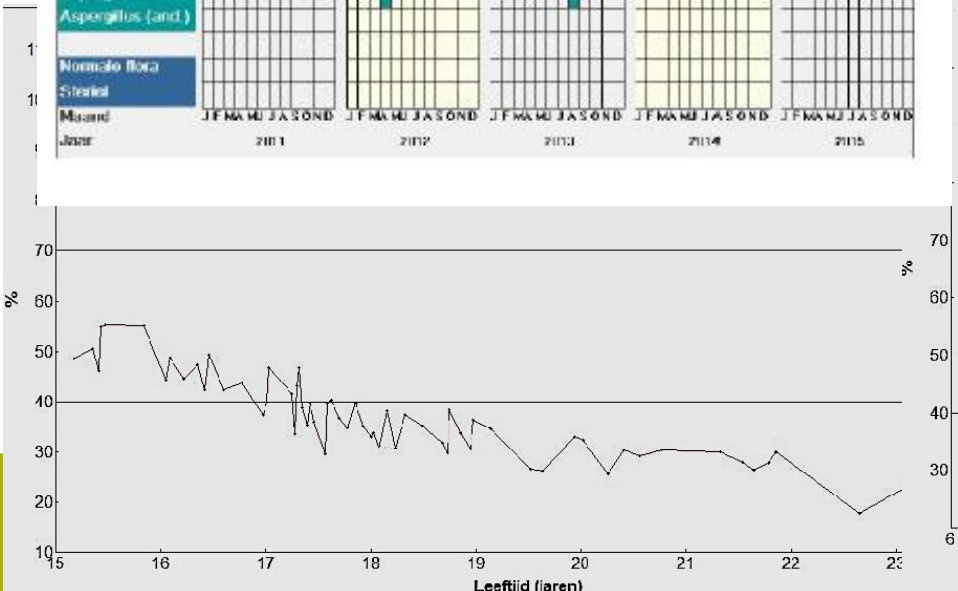


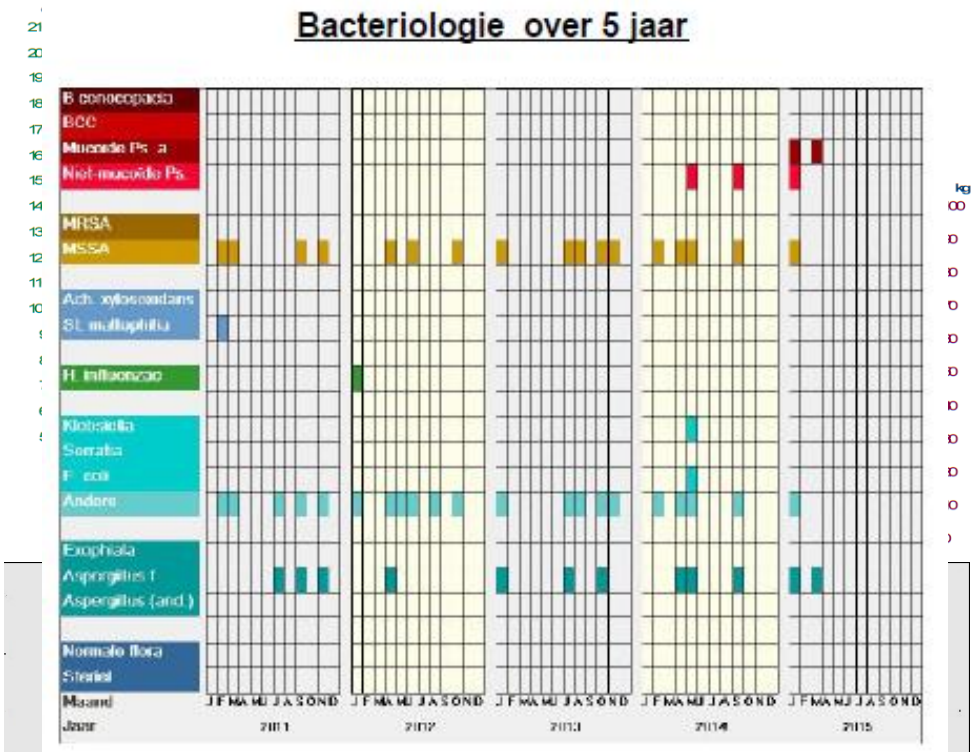


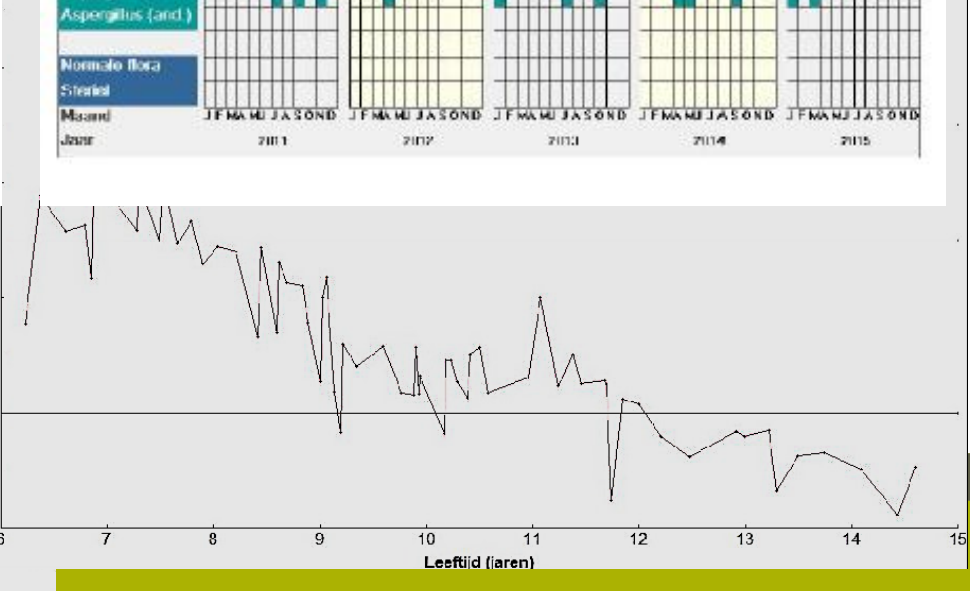


Leila °1992 Nadia °2000









Challenges

- -minimization of disease
- -decline despite severe treatment
- -explain why different course in both sisters
- -accept “invasive” procedures (portacath, gastrostomy…)
- -transplantation
- -accept patient’s demands/negotiations