Eosinophilic Fasciitis

the patient experience based on patient-organised web forums

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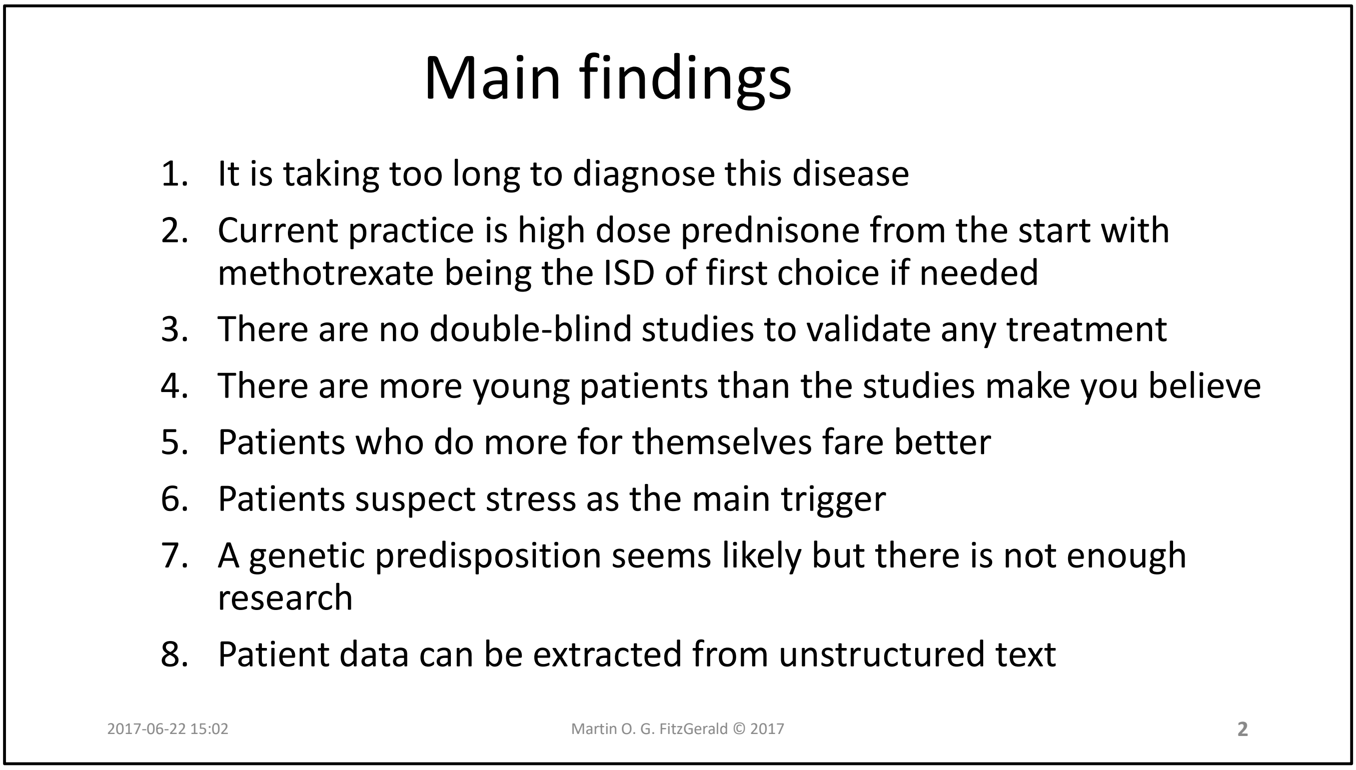
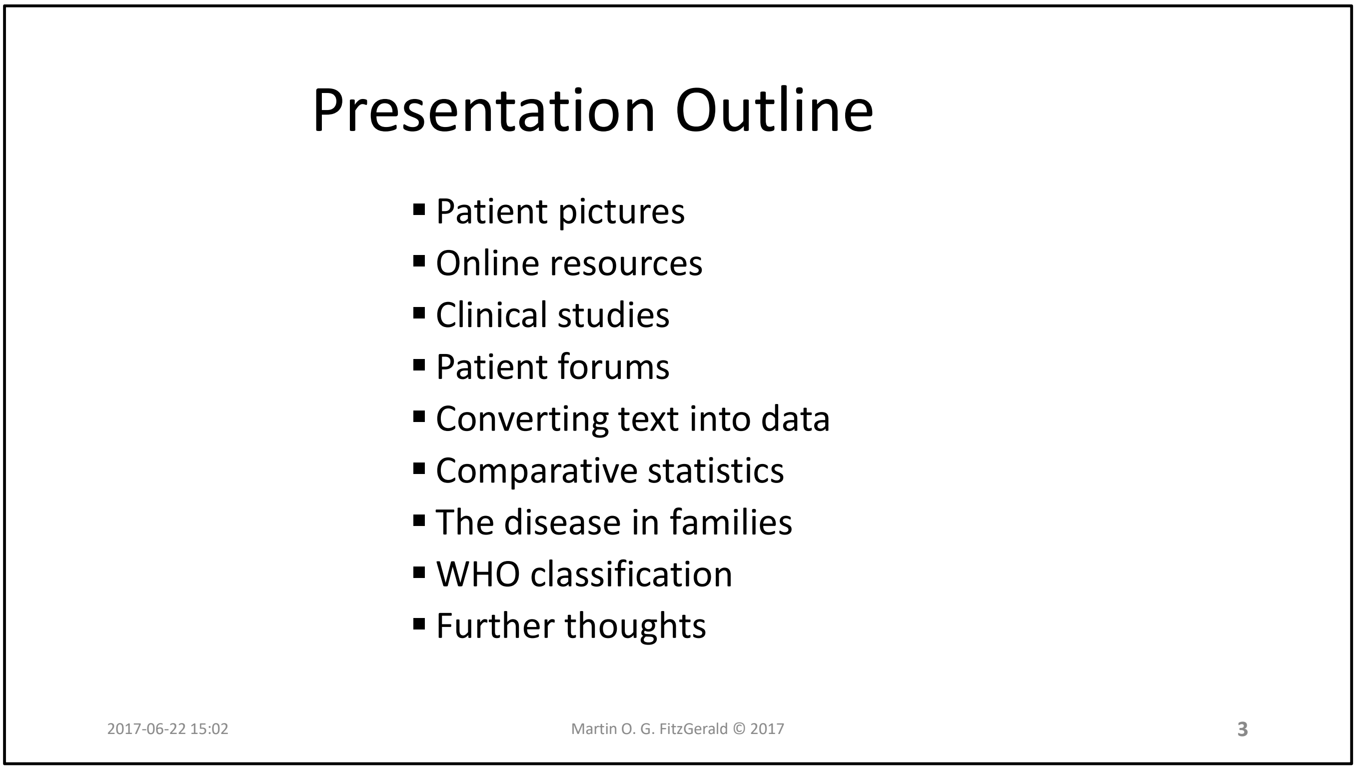
Thank you Professor Chizzolini for your introduction and the opportunity to talk today.
As you now know I am a patient with Eosinophilic Fasciitis whose disease is in remission.
What I want to share with you is drawn from my own experience and what other patients have posted in online forums. I will call this forum data.



Main findings

1. It is taking too long to diagnose this disease
2. Current practice is high dose prednisone from the start with methotrexate being the ISD of first choice if needed
3. There are no double-blind studies to validate any treatment
4. There are more young patients than the studies make you believe
5. Patients who do more for themselves fare better
6. Patients suspect stress as the main trigger
7. A genetic predisposition seems likely but there is not enough research
8. Patient data can be extracted from unstructured text

All these findings will be addressed by the slides.
(1) The forum data shows it is taking to long to diagnose this disease
(2) Recent studies show that this treatment is effective and the forum data generally support it [ISD=immuno-suppressant drug]
(3) There are too few patients for double-blind studies
(4) The forum data contains more young patients than the studies
(5) The forum statistics show that proactive patients do better
(6) The clinical studies propose unusual physical exertion as the most common trigger, but patients put more blame on stress
(7) There is some evidence for EF occurring in families
(8) What this project tries to demonstrate is how data mining of what patients **write**, can add to what doctors **know**



Presentation Outline

- Patient pictures
- Online resources
- Clinical studies
- Patient forums
- Converting text into data
- Comparative statistics
- The disease in families
- WHO classification
- Further thoughts

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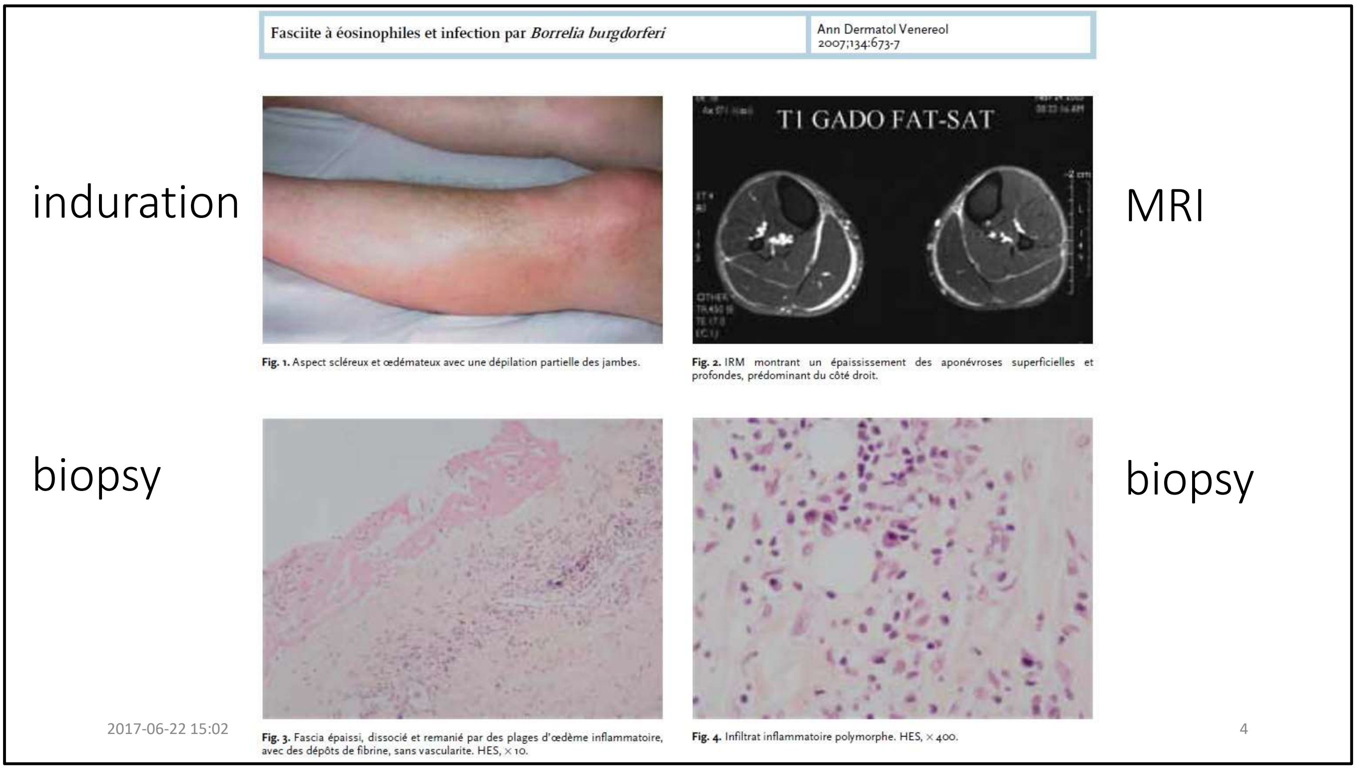
I will start with some pictures just to give an idea of this disease. Then see what patients find online. Refer to some studies. Show where patients post their stories. Show how we can convert that to data and run queries. See some cases of this disease in families. See how the W-H-O classifies this disease and finish up with some thoughts and suggestions.



Fasciite à éosinophiles et infection par *Borrelia burgdorferi*

Ann Dermatol Venereol
2007;134:673-7

induration

MRI

Fig. 1. Aspect scléreux et œdémateux avec une dépilation partielle des jambes.

Fig. 2. IRM montrant un épaississement des aponévroses superficielles et profondes, prédominant du côté droit.

biopsy

biopsy

Fig. 3. Fascia épaissi, dissocié et remanié par des plages d'œdème inflammatoire, avec des dépôts de fibrine, sans vascularite. HES, × 10.

Fig. 4. Infiltrat inflammatoire polymorphe. HES, × 400.

These pictures are from one of the studies. My legs looked like this. The tight shiny edematous skin is typical of this disease. The MRI was a turning point in my diagnosis. My generalist said he would not have been able to interpret my MRI pictures, only a specialist could. Perhaps there is software that can. The biopsy samples apparently show fascia inflammation and infiltration of eosinophils. To me they look just like modern art.

Figure 1. Infiltrated areas in both forearms.

Figure 2. Infiltrated area in right leg, sclerosis of skin, and scarcity of hair in the affected area.

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This shows how to check the Modified Rodnan Skin Score. I had this done each month to measure my progress.

FIG 1 Classical symptoms of eosinophilic fasciitis with venous furrowing on the arms of the patient.

I had grooves like this on both sides of my arms. When I showed an intern, unsurprisingly he had no idea what they were. I imagine these grooves are not a major topic at medical school.

Fig. 1. *Peau d'orange.*

Figure 1 Eosinophilic fasciitis related skin induration of the leg

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My arms had the peau d’orange look, like this leg. But not like that arm which is at a worse stage of the disease. That lumpy appearance seems to affect women more than men. One of my physiotherapists thought it might be an adipose effect.

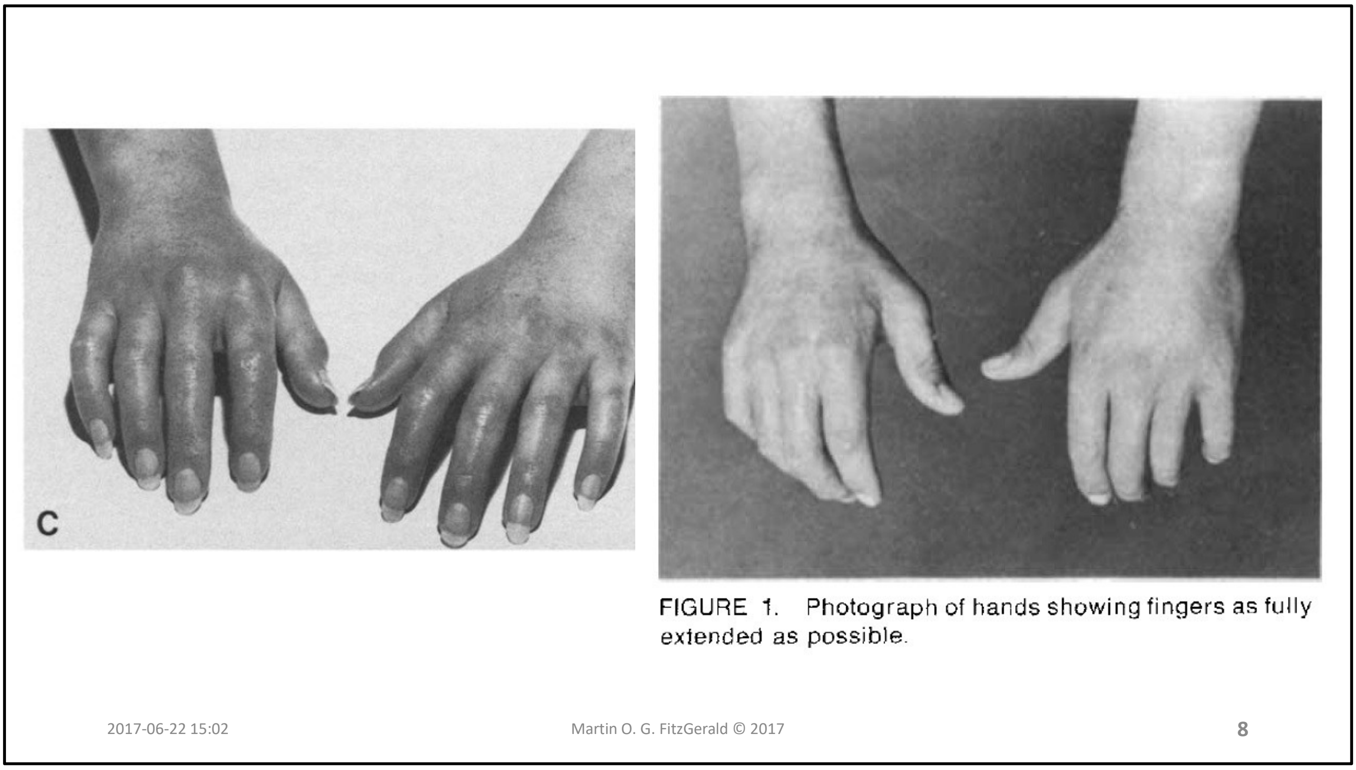

FIGURE 1. Photograph of hands showing fingers as fully extended as possible.

I had swollen hands at first which turned into the famous “claw hands” shown here. I couldn’t open or close them properly. It is a very common symptom of this disease and is often described as carpal tunnel syndrome but I am not sure if it’s really the same thing when the underlying cause is different.

What can patients find online about this disease?

I was alarmed by my symptoms and worried about getting worse. Like other patients, I looked for answers online.

Professional health sites for EF patients

http://www.merckmanuals.com/.../eosinophilic-fasciitis/

MERCK MANUAL Professional Version

https://rarediseases.org/rare-diseases/eosinophilic-fasciitis/

NORD® National Organization for Rare Disorders

http://apfed.org/about-ead/eosinophilic-fasciitis/

Apfed American Partnership for Eosinophilic Disorders

http://www.mdguidelines.com/eosinophilic-fasciitis

ReedGroup® MDGuidelines™

http://www.sclero.org/scleroderma/autoimmune/skin-diseases/eosinophilic-fasciitis/a-to-z.html

SCLERO.ORG

https://rarediseases.info.nih.gov/diseases/6351/index

GARD Genetic and Rare Diseases Information Center

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Although it’s still common advice to avoid looking up your condition online, in today’s digital world this is unavoidable. So what can patients find? Here are some patient-friendly sites about this disease which are informative and have no paywall.

What else is there?

The clinical studies.

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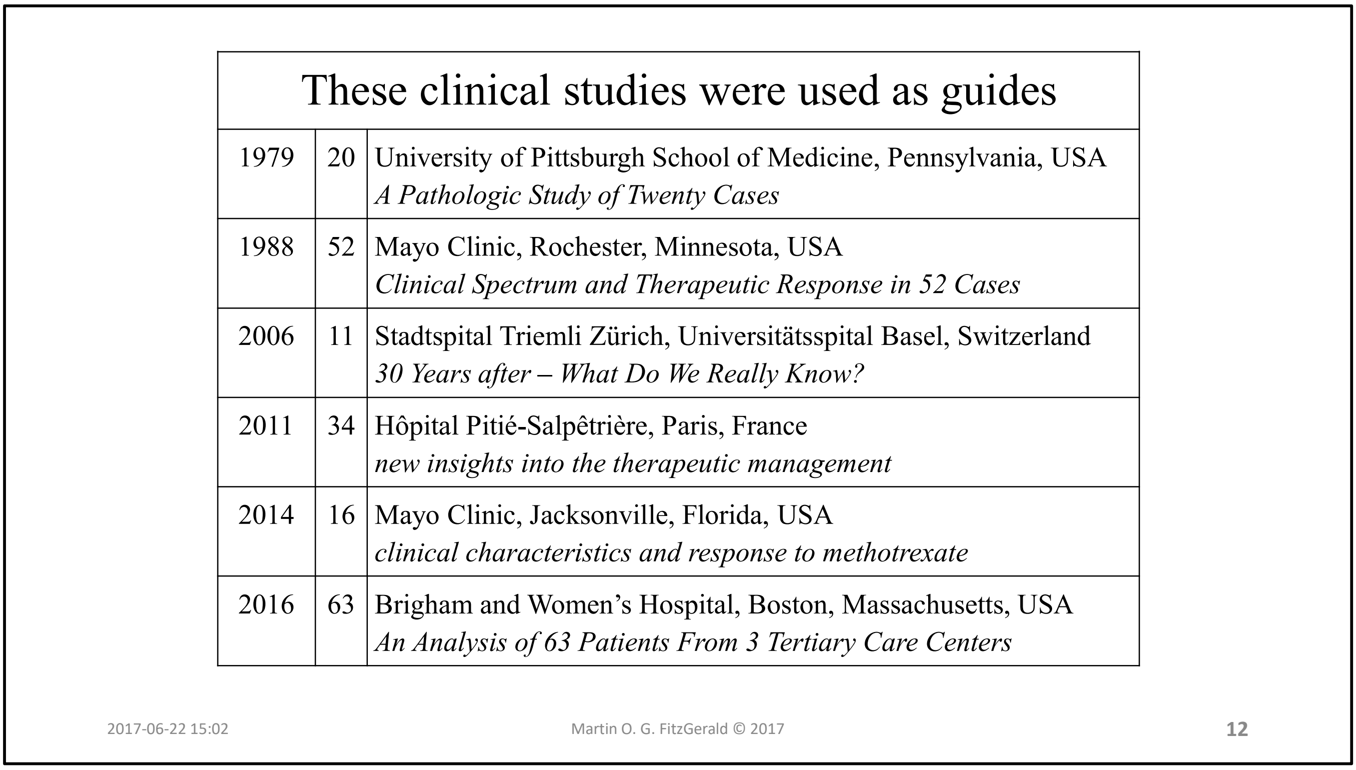
I learned about these from another patient who recommended them.



These clinical studies were used as guides

1979	20	University of Pittsburgh School of Medicine, Pennsylvania, USA *A Pathologic Study of Twenty Cases*
1988	52	Mayo Clinic, Rochester, Minnesota, USA *Clinical Spectrum and Therapeutic Response in 52 Cases*
2006	11	Stadtspital Triemli Zürich, Universitätsspital Basel, Switzerland *30 Years after – What Do We Really Know?*
2011	34	Hôpital Pitié-Salpêtrière, Paris, France *new insights into the therapeutic management*
2014	16	Mayo Clinic, Jacksonville, Florida, USA *clinical characteristics and response to methotrexate*
2016	63	Brigham and Women’s Hospital, Boston, Massachusetts, USA *An Analysis of 63 Patients From 3 Tertiary Care Centers*

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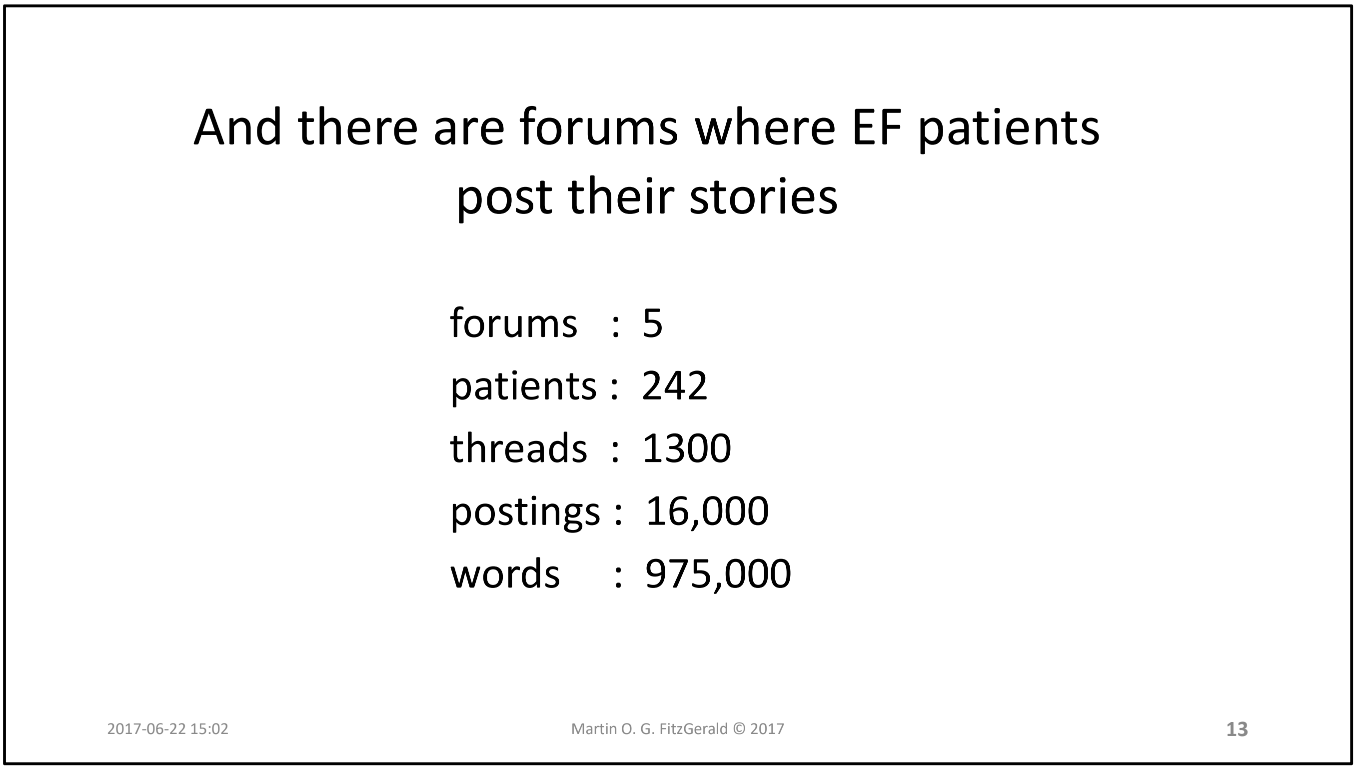
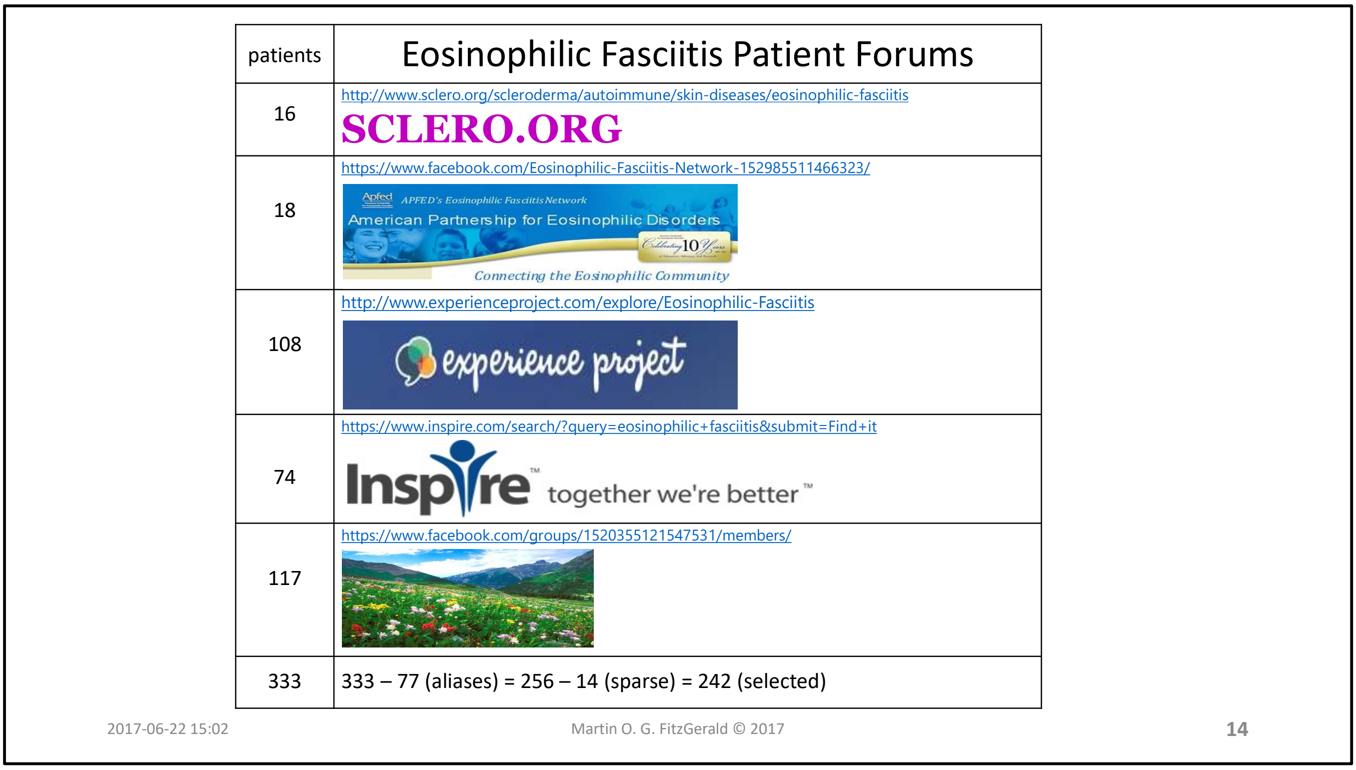
These studies are interesting for their fairly large patient cohorts. The 1988 Mayo Clinic study seemed to set a standard for the others. The 2016 Brigham and Women’s study found there was a strong case for the combination of corticosteroids and methotrexate. It also found that patients diagnosed within 6 months fared slightly better. I looked for this correlation among the forum patients but it wasn’t obvious. That seems counter-intuitive but I have to respect the data. So far there have been no double-blind studies to validate treatments. There are simply not enough of us. Perhaps there have been double-blind studies on other eosinophilic disorders that might be informative.



And there are forums where EF patients post their stories

forums : 5
patients : 242
threads : 1300
postings : 16,000
words : 975,000

This was the input to my project. I selected 242 patients for inclusion in the database. You can see that they posted nearly a million words, all of which had to be read and converted to data.



patients	Eosinophilic Fasciitis Patient Forums
16	http://www.sclero.org/scleroderma/autoimmune/skin-diseases/eosinophilic-fasciitis SCLERO.ORG
18	https://www.facebook.com/Eosinophilic-Fasciitis-Network-152985511466323/ APFED's Eosinophilic Fasciitis Network American Partnership for Eosinophilic Disorders Celebrating 10 Years Connecting the Eosinophilic Community
108	http://www.experienceproject.com/explore/Eosinophilic-Fasciitis experience project
74	https://www.inspire.com/search/?query=eosinophilic+fasciitis&submit=Find+it Inspire™ together we're better™
117	https://www.facebook.com/groups/1520355121547531/members/
333	333 – 77 (aliases) = 256 – 14 (sparse) = 242 (selected)

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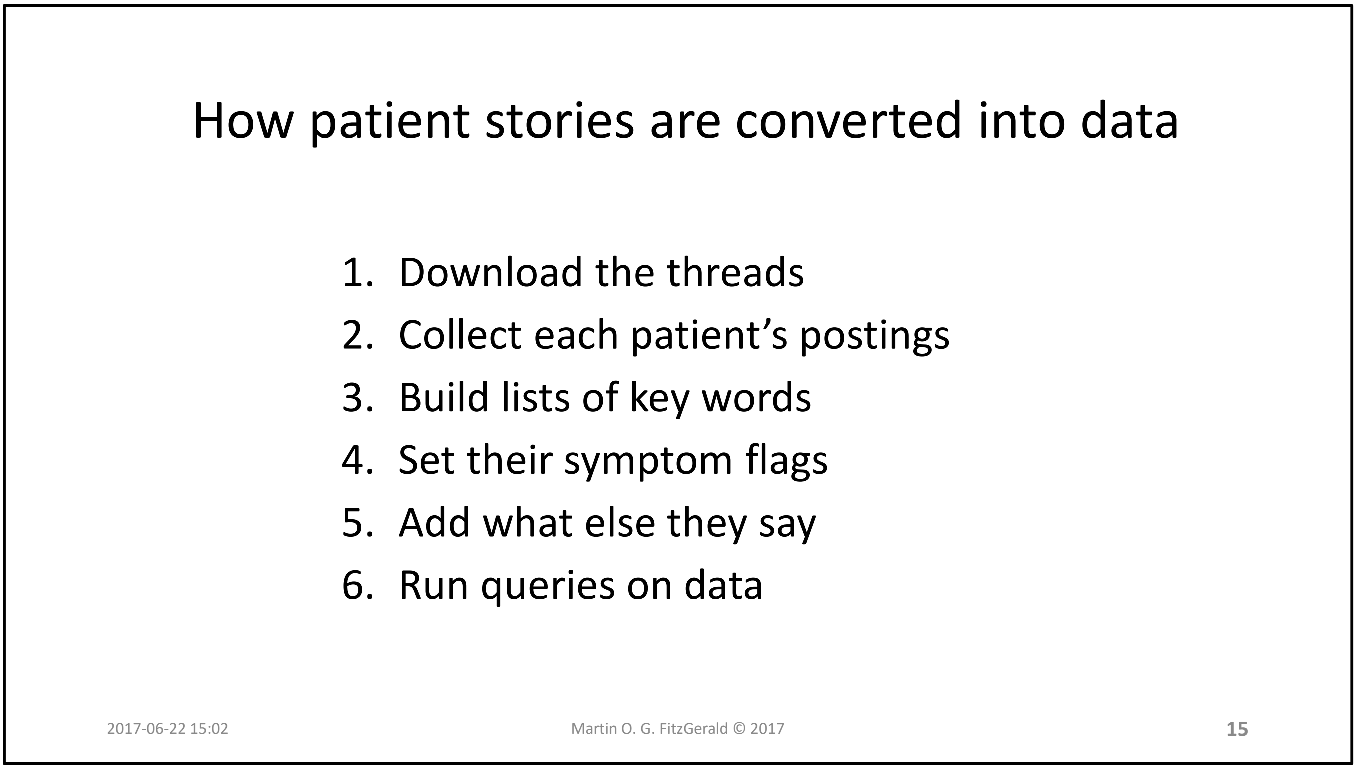
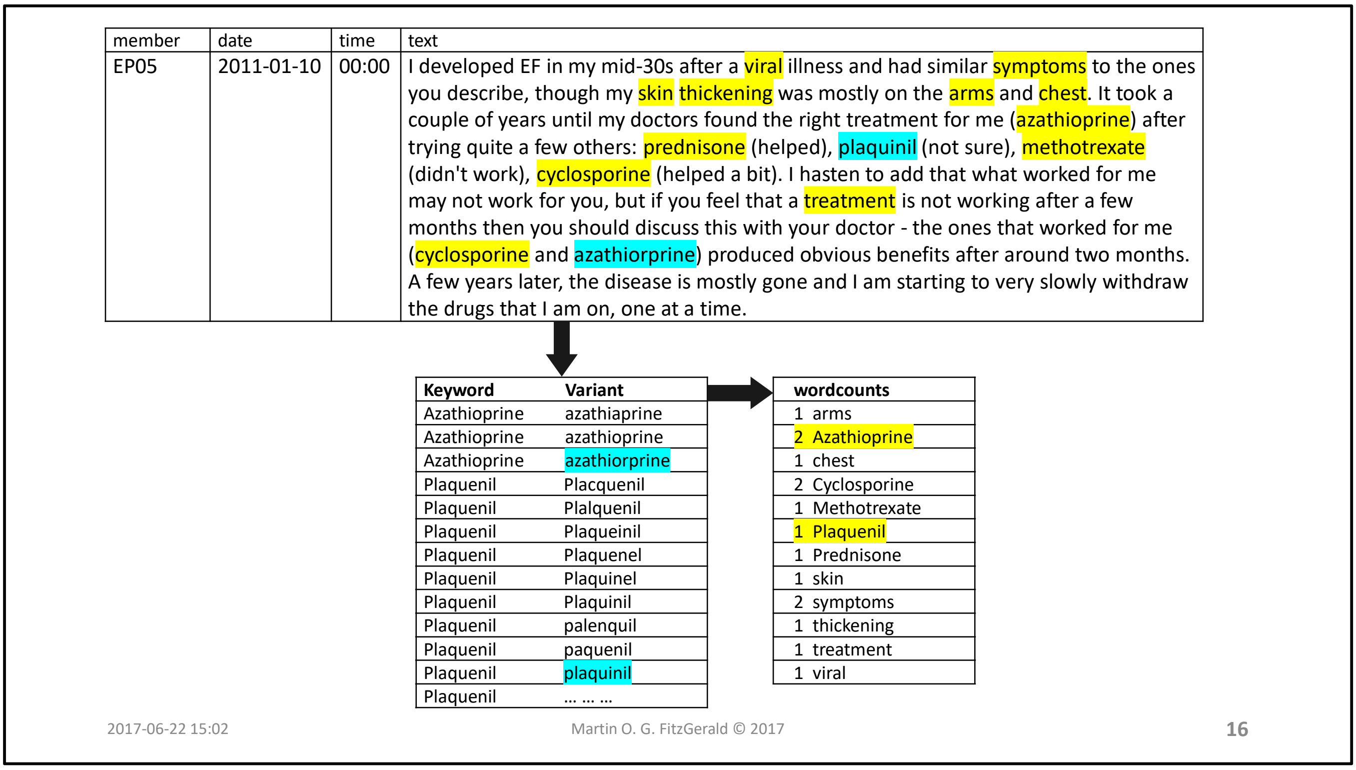
These are the places where patients posted their stories. There is moral support, no criticism, no time constraints and the comfort of being at home. Some patients are health professionals who give good advice. Most of the patients who post in these forums are from North America and English speaking countries, only a few are from Europe. The **global** occurrence of this disease is still unknown.



How patient stories are converted into data

1. Download the threads
2. Collect each patient’s postings
3. Build lists of key words
4. Set their symptom flags
5. Add what else they say
6. Run queries on data

I used my IT knowledge (I am a software developer) to format and load the patient data into a relational database. Once that was done it was easy to write queries that looked for interesting things.

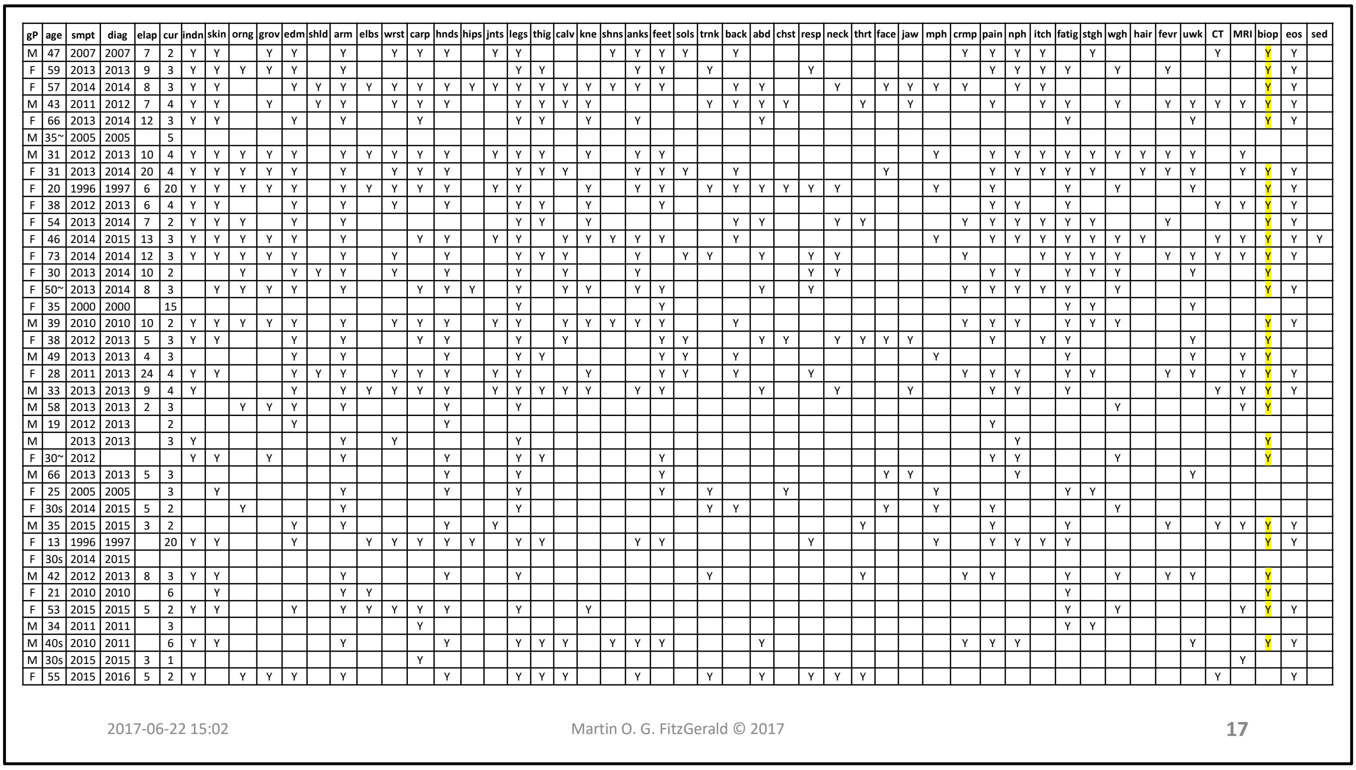


member	date	time	text
EP05	2011-01-10	00:00	I developed EF in my mid-30s after a viral illness and had similar symptoms to the ones you describe, though my skin thickening was mostly on the arms and chest. It took a couple of years until my doctors found the right treatment for me (azathioprine) after trying quite a few others: prednisone (helped), plaquinil (not sure), methotrexate (didn't work), cyclosporine (helped a bit). I hasten to add that what worked for me may not work for you, but if you feel that a treatment is not working after a few months then you should discuss this with your doctor - the ones that worked for me (cyclosporine and azathiorprine) produced obvious benefits after around two months. A few years later, the disease is mostly gone and I am starting to very slowly withdraw the drugs that I am on, one at a time.

Keyword	**Variant**
Azathioprine	azathiaprine
Azathioprine	azathioprine
Azathioprine	azathiorprine
Plaquenil	Placquenil
Plaquenil	Plalquenil
Plaquenil	Plaqueinil
Plaquenil	Plaquenel
Plaquenil	Plaquinel
Plaquenil	Plaquinil
Plaquenil	palenquil
Plaquenil	paquenil
Plaquenil	plaquinil
Plaquenil	… … …

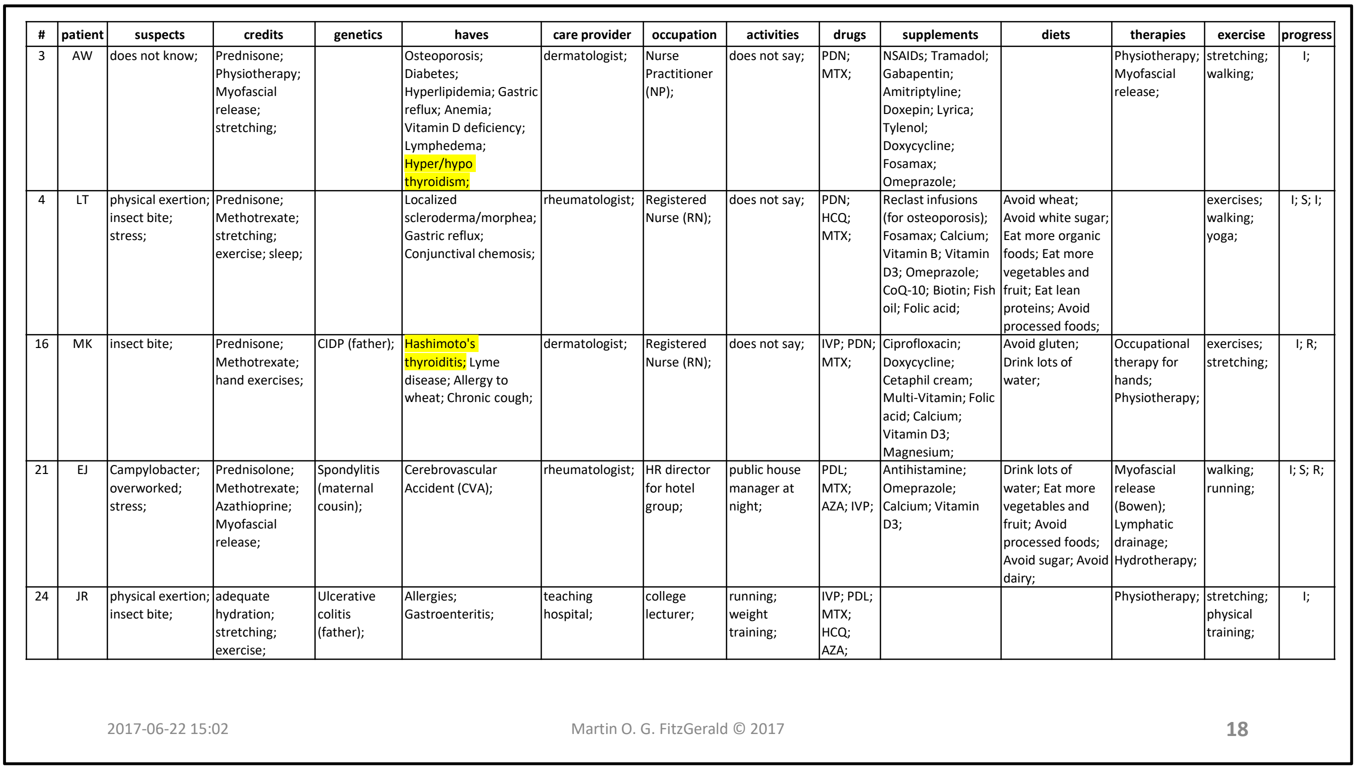
wordcounts
1 arms
2 Azathioprine
1 chest
2 Cyclosporine
1 Methotrexate
1 Plaquenil
1 Prednisone
1 skin
2 symptoms
1 thickening
1 treatment
1 viral

I could not automatically convert what patients say into data. I had to read it all. But I made this tool that extracts the "interesting" words for each patient. I did not make a "white list" of these words, but did the opposite. I made an exclusion list of the "uninteresting" words so that whatever words are left are selected and counted. This ensures that new words are interesting until I decide they are not. A lookup table handles misspellings.



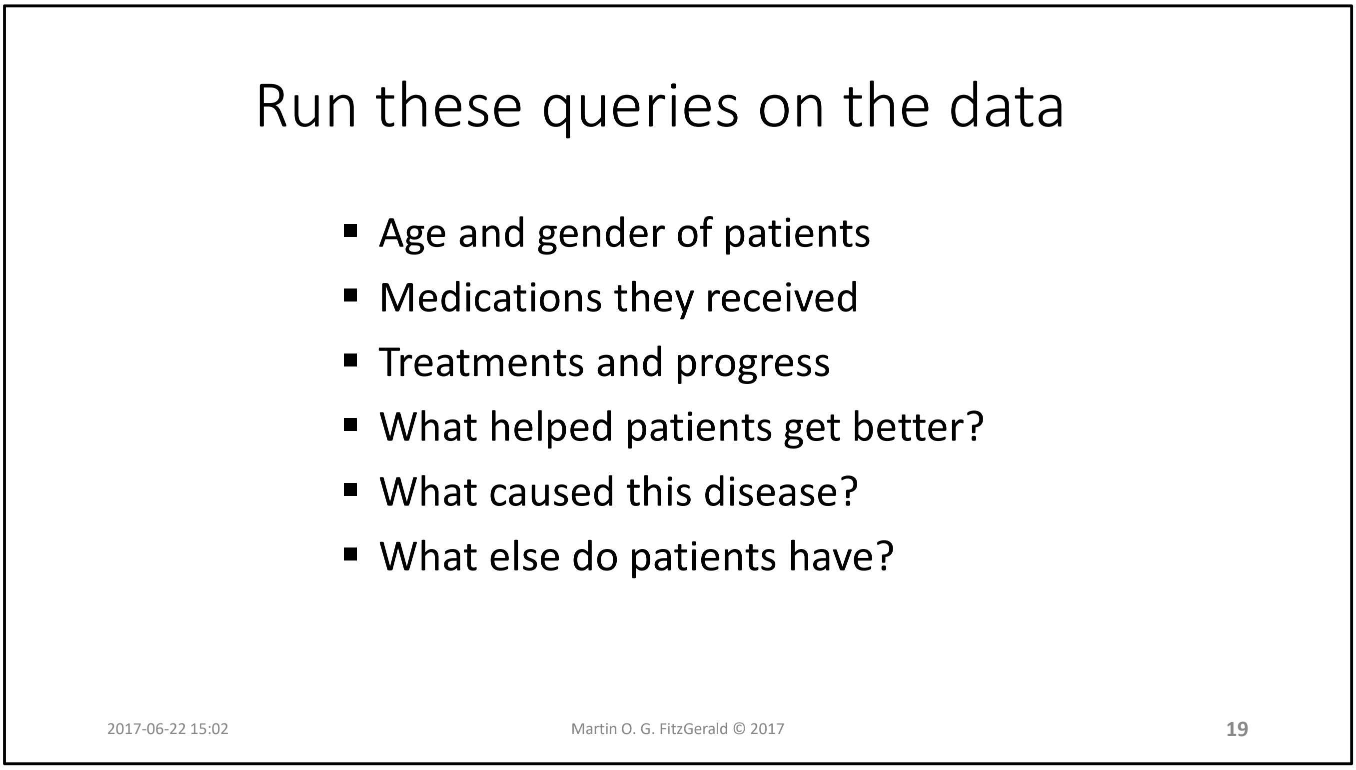
gP	age	smpt	diag	elap	cur	indn	skin	orng	grov	edm	shld	arm	elbs	wrst	carp	hnds	hips	jnts	legs	thig	calv	kne	shns	anks	feet	sols	trnk	back	abd	chst	resp	neck	thrt	face	jaw	mph	crmp	pain	nph	itch	fatig	stgh	wgh	hair	fevr	uwk	CT	MRI	biop	eos	sed
M	47	2007	2007	7	2	Y	Y		Y	Y		Y		Y	Y	Y		Y	Y				Y	Y	Y	Y		Y									Y	Y	Y	Y		Y					Y		Y	Y	
F	59	2013	2013	9	3	Y	Y	Y	Y	Y		Y							Y	Y				Y	Y		Y				Y							Y	Y	Y	Y		Y		Y				Y	Y	
F	57	2014	2014	8	3	Y	Y			Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y			Y	Y			Y		Y	Y	Y	Y		Y	Y									Y	Y	
M	43	2011	2012	7	4	Y	Y		Y		Y	Y		Y	Y	Y			Y	Y	Y	Y					Y	Y	Y	Y			Y		Y			Y		Y	Y		Y		Y	Y	Y	Y	Y	Y	
F	66	2013	2014	12	3	Y	Y			Y		Y			Y				Y	Y		Y		Y					Y												Y					Y			Y	Y	
M	35~	2005	2005		5																																														
M	31	2012	2013	10	4	Y	Y	Y	Y	Y		Y	Y	Y	Y	Y		Y	Y	Y		Y		Y	Y											Y		Y	Y	Y	Y	Y	Y	Y	Y	Y		Y			
F	31	2013	2014	20	4	Y	Y	Y	Y	Y		Y		Y	Y	Y			Y	Y	Y			Y	Y	Y		Y						Y				Y	Y	Y	Y	Y		Y	Y	Y		Y	Y	Y	
F	20	1996	1997	6	20	Y	Y	Y	Y	Y		Y	Y	Y	Y	Y		Y	Y			Y		Y	Y		Y	Y	Y	Y	Y	Y				Y		Y			Y		Y			Y			Y	Y	
F	38	2012	2013	6	4	Y	Y			Y		Y		Y		Y			Y	Y		Y			Y													Y	Y		Y						Y	Y	Y	Y	
F	54	2013	2014	7	2	Y	Y	Y		Y		Y							Y	Y		Y						Y	Y			Y	Y				Y	Y	Y	Y	Y	Y			Y				Y	Y	
F	46	2014	2015	13	3	Y	Y	Y	Y	Y		Y			Y	Y		Y	Y		Y	Y	Y	Y	Y			Y								Y		Y	Y	Y	Y	Y	Y	Y			Y	Y	Y	Y	Y
F	73	2014	2014	12	3	Y	Y	Y	Y	Y		Y		Y		Y			Y	Y	Y			Y		Y	Y		Y		Y	Y					Y			Y	Y	Y	Y		Y	Y	Y	Y	Y	Y	
F	30	2013	2014	10	2			Y		Y	Y	Y		Y		Y			Y		Y			Y							Y	Y						Y	Y		Y	Y	Y			Y			Y		
F	50~	2013	2014	8	3		Y	Y	Y	Y		Y			Y	Y	Y		Y		Y	Y		Y	Y				Y		Y						Y	Y	Y	Y	Y		Y						Y	Y	
F	35	2000	2000		15														Y						Y																Y	Y				Y					
M	39	2010	2010	10	2	Y	Y	Y	Y	Y		Y		Y	Y	Y		Y	Y		Y	Y	Y	Y	Y			Y									Y	Y	Y		Y	Y	Y						Y	Y	
F	38	2012	2013	5	3	Y	Y			Y		Y			Y	Y			Y		Y				Y	Y			Y	Y		Y	Y	Y	Y			Y		Y	Y					Y			Y		
M	49	2013	2013	4	3					Y		Y				Y			Y	Y					Y	Y		Y								Y					Y					Y		Y	Y		
F	28	2011	2013	24	4	Y	Y			Y	Y	Y		Y	Y	Y		Y	Y			Y			Y	Y		Y			Y						Y	Y	Y		Y	Y			Y	Y		Y	Y	Y	
M	33	2013	2013	9	4	Y				Y		Y	Y	Y	Y	Y		Y	Y	Y	Y	Y		Y	Y				Y			Y			Y			Y	Y		Y						Y	Y	Y	Y	
M	58	2013	2013	2	3			Y	Y	Y		Y				Y			Y																								Y					Y	Y		
M	19	2012	2013		2					Y						Y																						Y													
M		2013	2013		3	Y						Y		Y					Y																				Y										Y		
F	30~	2012				Y	Y		Y			Y				Y			Y	Y					Y													Y	Y				Y						Y		
M	66	2013	2013	5	3											Y			Y						Y									Y	Y				Y							Y					
F	25	2005	2005		3		Y					Y				Y			Y						Y		Y			Y						Y					Y	Y									
F	30s	2014	2015	5	2			Y				Y							Y								Y	Y						Y		Y		Y					Y								
M	35	2015	2015	3	2					Y		Y				Y		Y															Y					Y			Y				Y		Y	Y	Y	Y	
F	13	1996	1997		20	Y	Y			Y			Y	Y	Y	Y	Y		Y	Y				Y	Y						Y					Y		Y	Y	Y	Y								Y	Y	
F	30s	2014	2015																																																
M	42	2012	2013	8	3	Y	Y					Y				Y			Y								Y						Y				Y	Y			Y		Y		Y	Y			Y		
F	21	2010	2010		6		Y					Y	Y																												Y								Y		
F	53	2015	2015	5	2	Y	Y			Y		Y	Y	Y	Y	Y			Y			Y																			Y		Y					Y	Y	Y	
M	34	2011	2011		3										Y																										Y	Y									
M	40s	2010	2011		6	Y	Y					Y				Y			Y	Y	Y		Y	Y	Y				Y								Y	Y	Y							Y			Y	Y	
M	30s	2015	2015	3	1										Y																																	Y			
F	55	2015	2016	5	2	Y		Y	Y	Y		Y				Y			Y	Y	Y			Y			Y		Y		Y	Y	Y														Y			Y	

This is just to give you an impression of the work. I identified 43 symptoms that patients complain about. The last 5 columns are related to their diagnosis. You can see how many mentioned a biopsy.



#	patient	suspects	credits	genetics	haves	care provider	occupation	activities	drugs	supplements	diets	therapies	exercise	progress
3	AW	does not know;	Prednisone; Physiotherapy; Myofascial release; stretching;		Osteoporosis; Diabetes; Hyperlipidemia; Gastric reflux; Anemia; Vitamin D deficiency; Lymphedema; Hyper/hypo thyroidism;	dermatologist;	Nurse Practitioner (NP);	does not say;	PDN; MTX;	NSAIDs; Tramadol; Gabapentin; Amitriptyline; Doxepin; Lyrica; Tylenol; Doxycycline; Fosamax; Omeprazole;		Physiotherapy; Myofascial release;	stretching; walking;	I;
4	LT	physical exertion; insect bite; stress;	Prednisone; Methotrexate; stretching; exercise; sleep;		Localized scleroderma/morphea; Gastric reflux; Conjunctival chemosis;	rheumatologist;	Registered Nurse (RN);	does not say;	PDN; HCQ; MTX;	Reclast infusions (for osteoporosis); Fosamax; Calcium; Vitamin B; Vitamin D3; Omeprazole; CoQ-10; Biotin; Fish oil; Folic acid;	Avoid wheat; Avoid white sugar; Eat more organic foods; Eat more vegetables and fruit; Eat lean proteins; Avoid processed foods;		exercises; walking; yoga;	I; S; I;
16	MK	insect bite;	Prednisone; Methotrexate; hand exercises;	CIDP (father);	Hashimoto's thyroiditis; Lyme disease; Allergy to wheat; Chronic cough;	dermatologist;	Registered Nurse (RN);	does not say;	IVP; PDN; MTX;	Ciprofloxacin; Doxycycline; Cetaphil cream; Multi-Vitamin; Folic acid; Calcium; Vitamin D3; Magnesium;	Avoid gluten; Drink lots of water;	Occupational therapy for hands; Physiotherapy;	exercises; stretching;	I; R;
21	EJ	Campylobacter; overworked; stress;	Prednisolone; Methotrexate; Azathioprine; Myofascial release;	Spondylitis (maternal cousin);	Cerebrovascular Accident (CVA);	rheumatologist;	HR director for hotel group;	public house manager at night;	PDL; MTX; AZA; IVP;	Antihistamine; Omeprazole; Calcium; Vitamin D3;	Drink lots of water; Eat more vegetables and fruit; Avoid processed foods; Avoid sugar; Avoid dairy;	Myofascial release (Bowen); Lymphatic drainage; Hydrotherapy;	walking; running;	I; S; R;
24	JR	physical exertion; insect bite;	adequate hydration; stretching; exercise;	Ulcerative colitis (father);	Allergies; Gastroenteritis;	teaching hospital;	college lecturer;	running; weight training;	IVP; PDL; MTX; HCQ; AZA;			Physiotherapy;	stretching; physical training;	I;

Everything else a patient says is grouped under 13 subjects to complete the patient record. The genetics column is for anything the patient has in their family. The haves column is for other conditions the patient has. I am interested in thyroid issues simply because my mother’s mother had Graves’ disease. She died during the thyroidectomy operation. My mother had systemic lupus. My father didn’t know what she had until a few weeks before she died, they didn’t tell him even though he was a doctor. That was in 1954. You would think that much has changed since then, that doctors would include patients earlier in their deliberations. But from what the forum patients are saying and my own experience, this is not happening as much as it should. During the weeks leading up to my diagnosis there were times when I felt like a bystander instead of the owner of my disease.



Run these queries on the data

- Age and gender of patients
- Medications they received
- Treatments and progress
- What helped patients get better?
- What caused this disease?
- What else do patients have?

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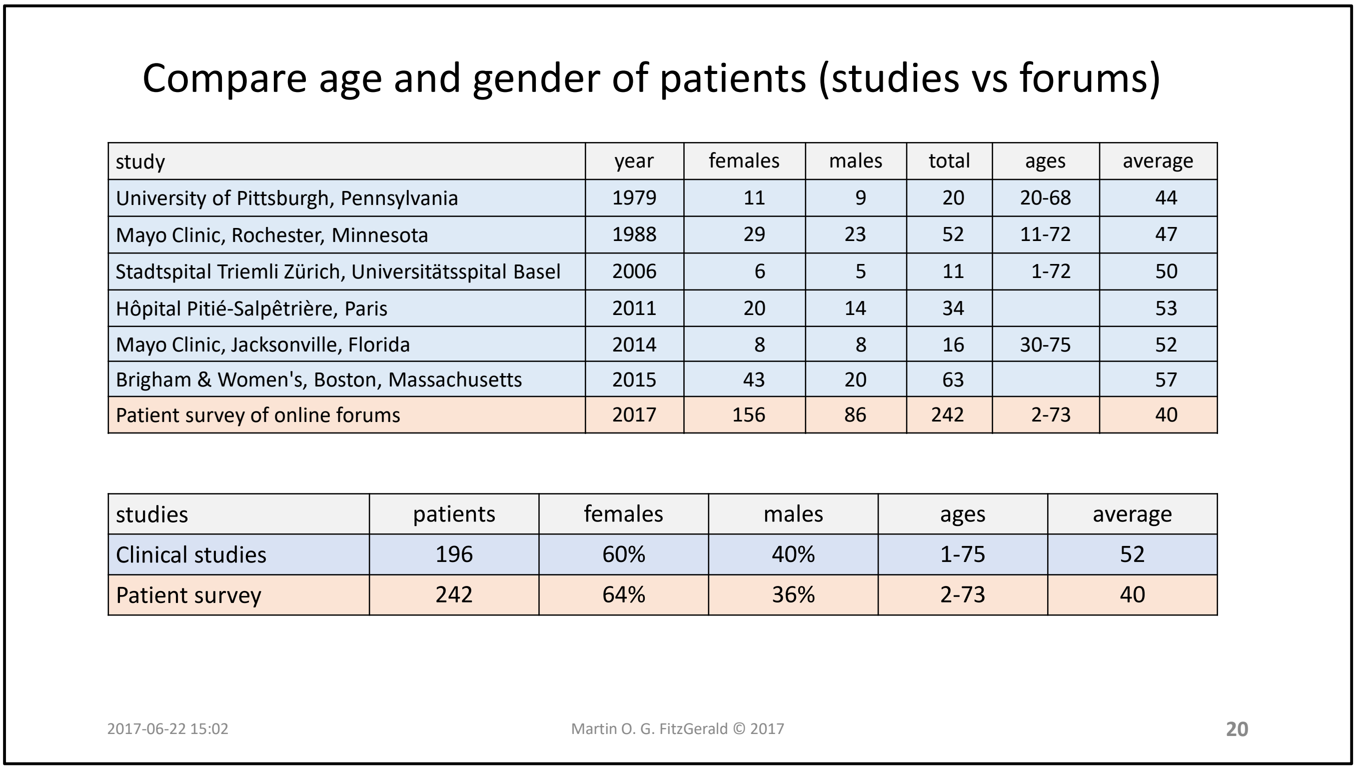
I wanted to see how the forum patients compared with the clinical studies. The next few slides show where they agree and where they differ.

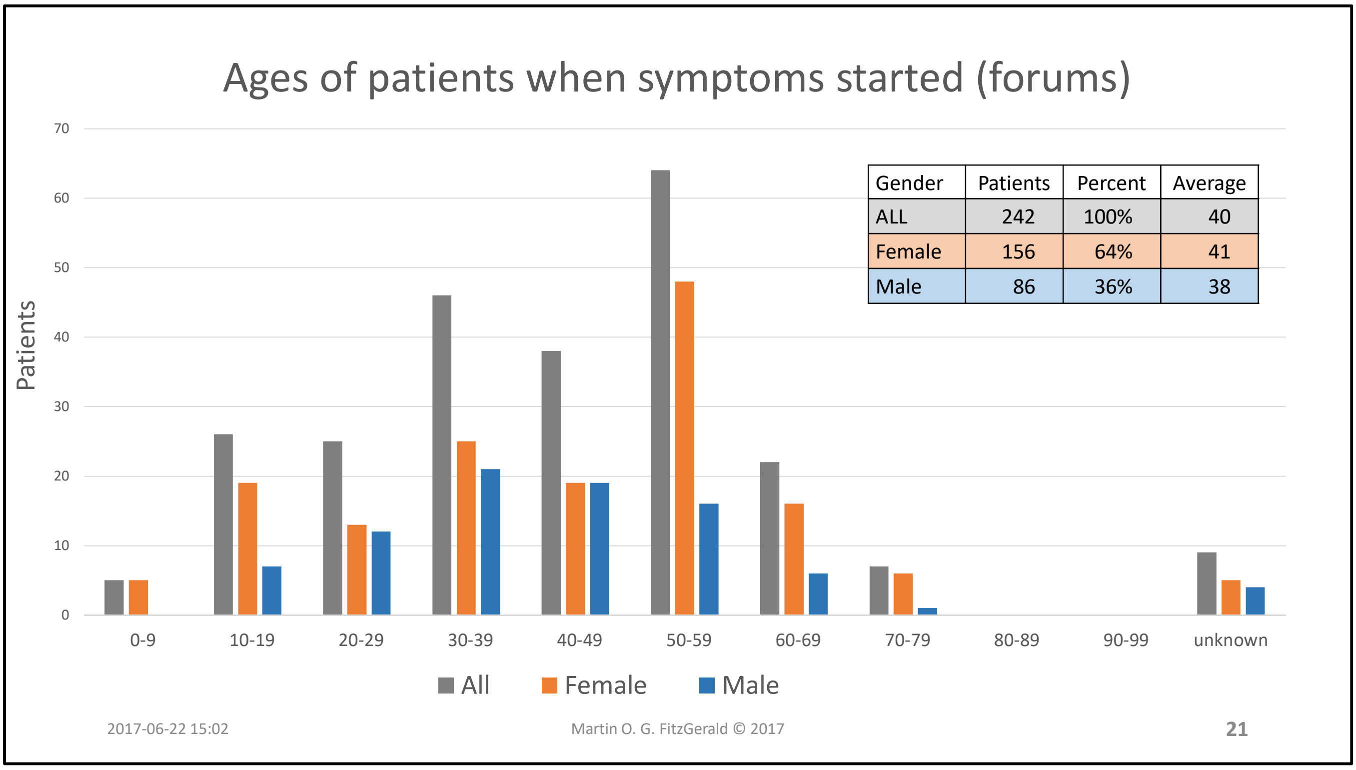


Compare age and gender of patients (studies vs forums)

study	year	females	males	total	ages	average
University of Pittsburgh, Pennsylvania	1979	11	9	20	20-68	44
Mayo Clinic, Rochester, Minnesota	1988	29	23	52	11-72	47
Stadtspital Triemli Zürich, Universitätsspital Basel	2006	6	5	11	1-72	50
Hôpital Pitié-Salpêtrière, Paris	2011	20	14	34		53
Mayo Clinic, Jacksonville, Florida	2014	8	8	16	30-75	52
Brigham & Women's, Boston, Massachusetts	2015	43	20	63		57
Patient survey of online forums	2017	156	86	242	2-73	40

studies	patients	females	males	ages	average
Clinical studies	196	60%	40%	1-75	52
Patient survey	242	64%	36%	2-73	40

After summarizing the clinical studies in one line we can see that forum patients appear on average to be younger than the studies have suggested. It is also interesting that the gender balance almost matches the clinical studies, this was unexpected as I had heard that females far outnumber males in online health forums.



Ages of patients when symptoms started (forums)

Gender	Patients	Percent	Average
ALL	242	100%	40
Female	156	64%	41
Male	86	36%	38

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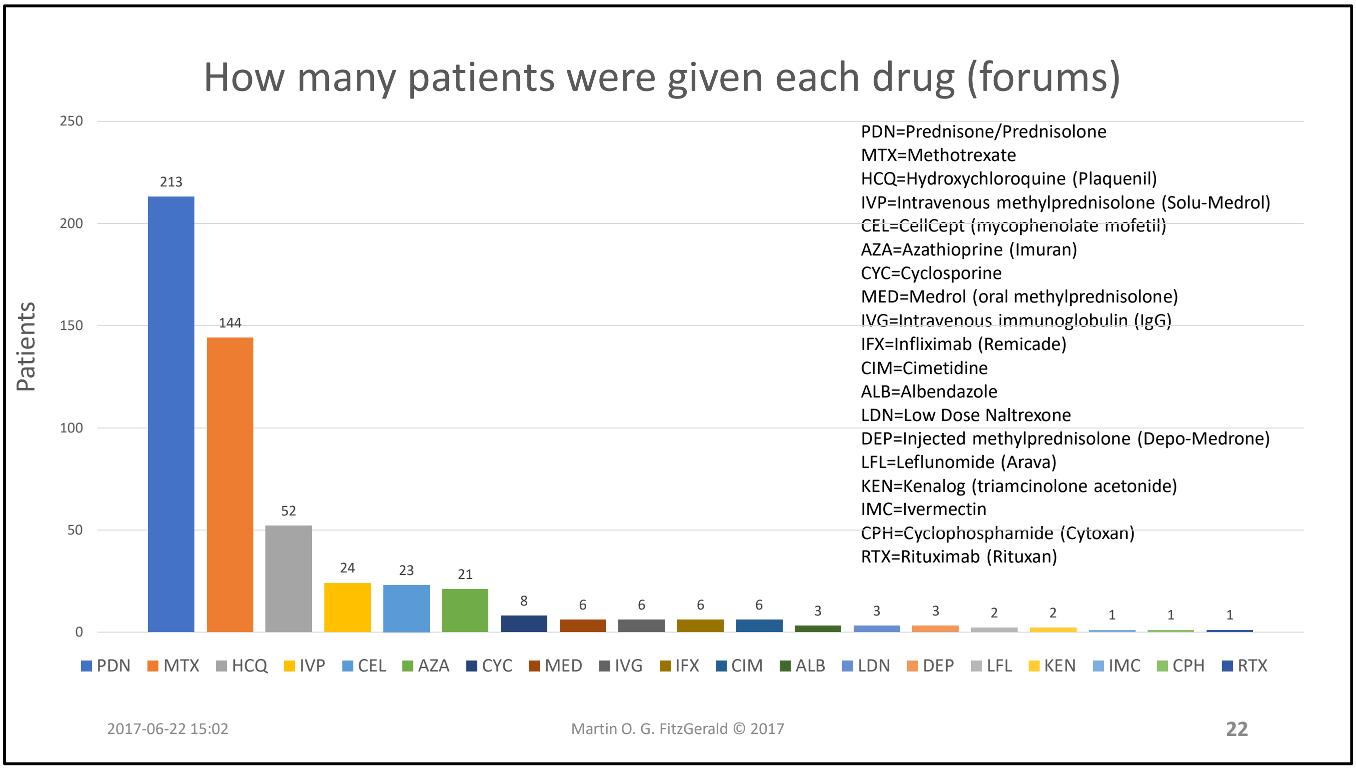
In age ranges, the forum data agree with the studies in that younger EF patients are mostly female. The peak of women in their 50s is a departure from the studies. It might be a forum effect. I just don’t know. You can see that the pattern for males is quite regular.



How many patients were given each drug (forums)

PDN=Prednisone/Prednisolone
MTX=Methotrexate
HCQ=Hydroxychloroquine (Plaquenil)
IVP=Intravenous methylprednisolone (Solu-Medrol)
CEL=CellCept (mycophenolate mofetil)
AZA=Azathioprine (Imuran)
CYC=Cyclosporine
MED=Medrol (oral methylprednisolone)
IVG=Intravenous immunoglobulin (IgG)
IFX=Infliximab (Remicade)
CIM=Cimetidine
ALB=Albendazole
LDN=Low Dose Naltrexone
DEP=Injected methylprednisolone (Depo-Medrone)
LFL=Leflunomide (Arava)
KEN=Kenalog (triamcinolone acetonide)
IMC=Ivermectin
CPH=Cyclophosphamide (Cytoxan)
RTX=Rituximab (Rituxan)

The less frequent drugs were usually prescribed only after the main ones had failed, but not always for that reason. Other health conditions sometimes limited the choices. I have kept the list of drugs to only those which were prescribed for Eosinophilic Fasciitis, not for other conditions the patients might have had.

Compare treatments and last known states

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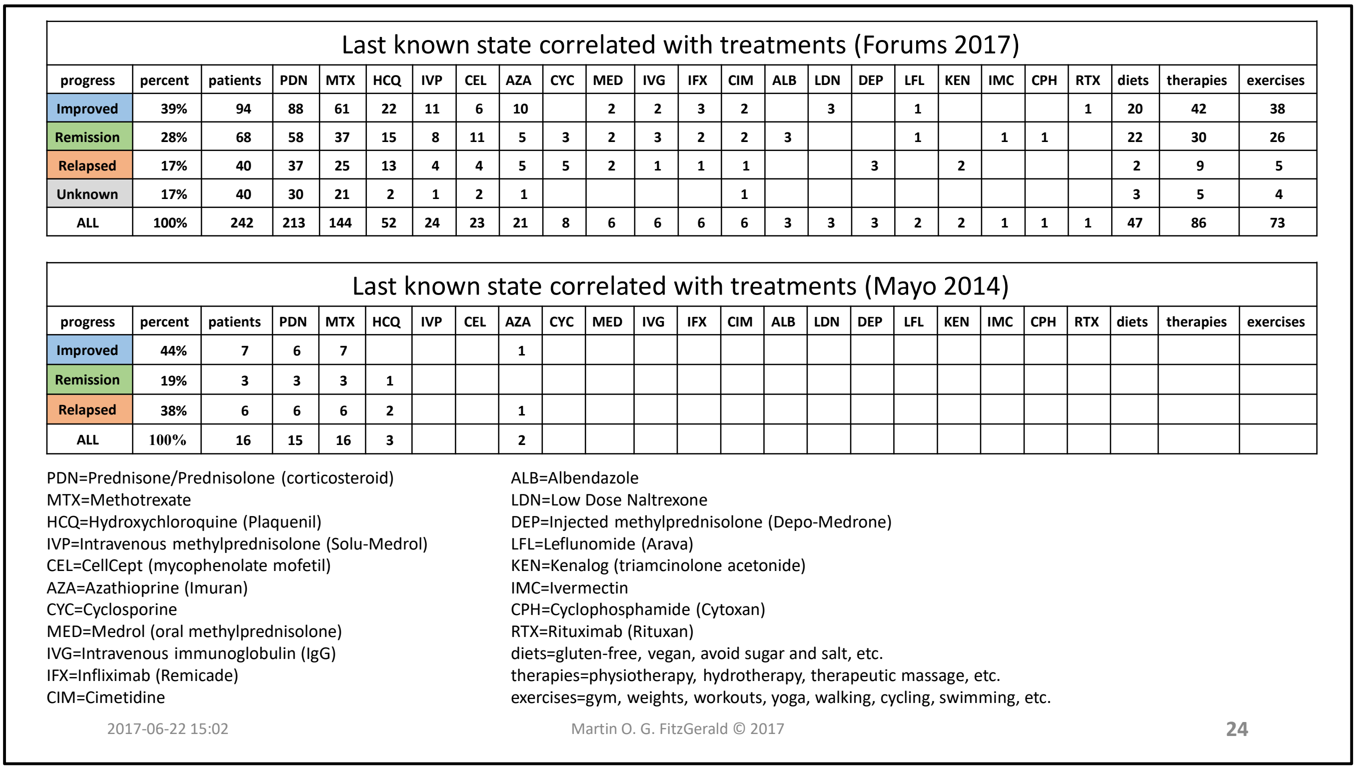
The next slide tries to correlate progress with treatments.



Last known state correlated with treatments (Forums 2017)

progress	percent	patients	PDN	MTX	HCQ	IVP	CEL	AZA	CYC	MED	IVG	IFX	CIM	ALB	LDN	DEP	LFL	KEN	IMC	CPH	RTX	diets	therapies	exercises
Improved	39%	94	88	61	22	11	6	10		2	2	3	2		3		1				1	20	42	38
Remission	28%	68	58	37	15	8	11	5	3	2	3	2	2	3			1		1	1		22	30	26
Relapsed	17%	40	37	25	13	4	4	5	5	2	1	1	1			3		2				2	9	5
Unknown	17%	40	30	21	2	1	2	1					1									3	5	4
ALL	100%	242	213	144	52	24	23	21	8	6	6	6	6	3	3	3	2	2	1	1	1	47	86	73

Last known state correlated with treatments (Mayo 2014)

progress	percent	patients	PDN	MTX	HCQ	IVP	CEL	AZA	CYC	MED	IVG	IFX	CIM	ALB	LDN	DEP	LFL	KEN	IMC	CPH	RTX	diets	therapies	exercises
Improved	44%	7	6	7				1																
Remission	19%	3	3	3	1																			
Relapsed	38%	6	6	6	2			1																
ALL	100%	16	15	16	3			2																

PDN=Prednisone/Prednisolone (corticosteroid)
MTX=Methotrexate
HCQ=Hydroxychloroquine (Plaquenil)
IVP=Intravenous methylprednisolone (Solu-Medrol)
CEL=CellCept (mycophenolate mofetil)
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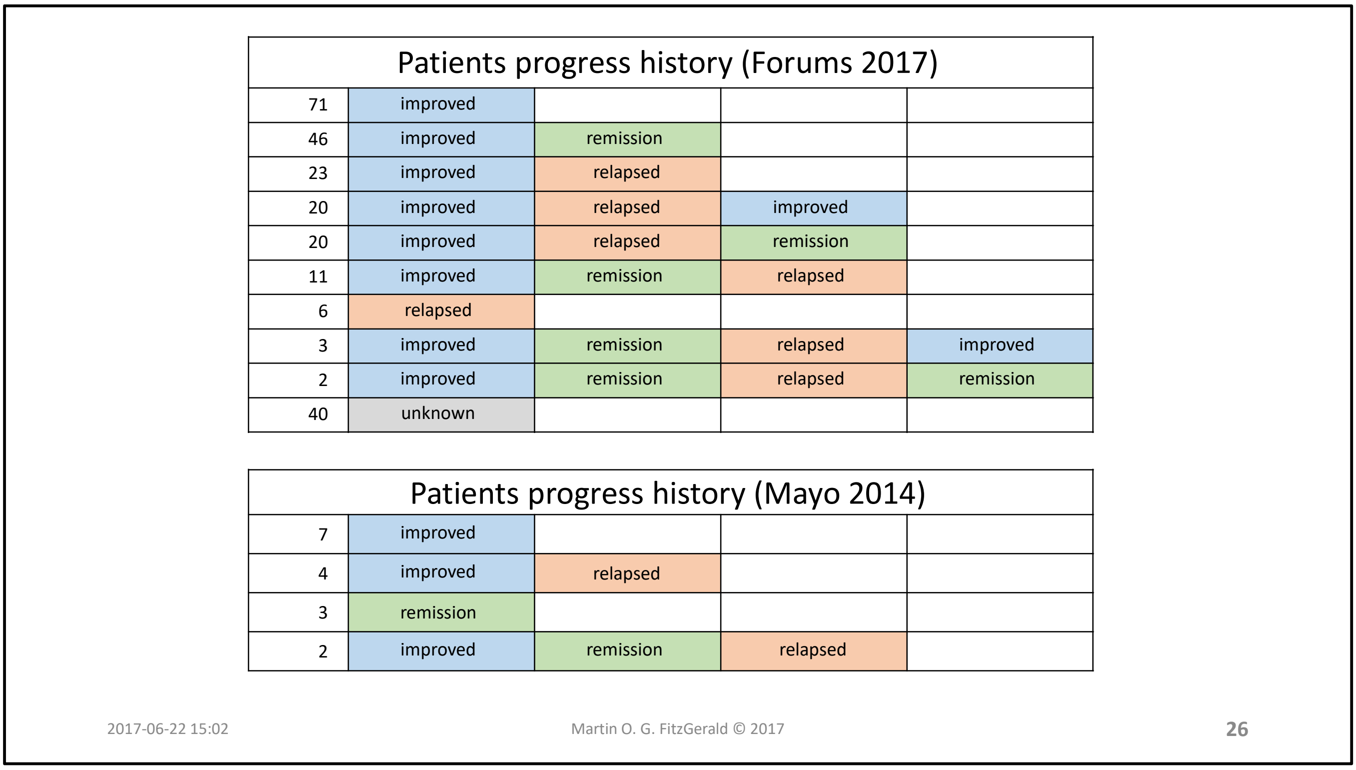
IMC=Ivermectin
CPH=Cyclophosphamide (Cytoxan)
RTX=Rituximab (Rituxan)
diets=gluten-free, vegan, avoid sugar and salt, etc.
therapies=physiotherapy, hydrotherapy, therapeutic massage, etc.
exercises=gym, weights, workouts, yoga, walking, cycling, swimming, etc.

It shows that the patients whose state is improved, or in remission, were more pro-active. This is an important finding that has come out of the patient stories. The Mayo 2014 study is shown just for comparison. Like most of the studies, it seems only interested in the drugs. I found some anomalies in their report which I have sent to them for comment.

How many patients relapsed?



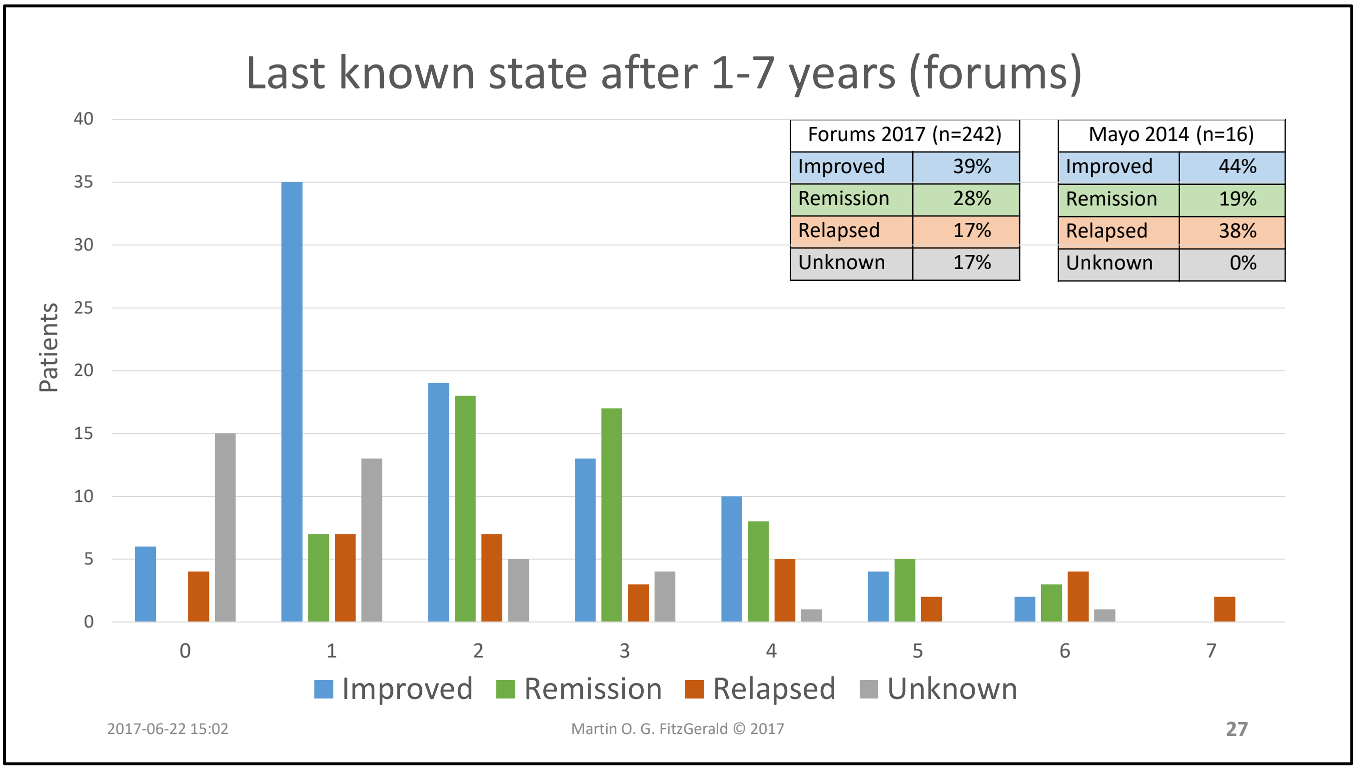
Patients progress history (Forums 2017)

71	improved			
46	improved	remission		
23	improved	relapsed		
20	improved	relapsed	improved	
20	improved	relapsed	remission	
11	improved	remission	relapsed	
6	relapsed			
3	improved	remission	relapsed	improved
2	improved	remission	relapsed	remission
40	unknown			

Patients progress history (Mayo 2014)

7	improved			
4	improved	relapsed		
3	remission			
2	improved	remission	relapsed	

In the forums about one third relapsed at some point. A few patients go into a remission-relapse-remission cycle that lasts for years. Some say they can control it with diet and exercise. The Mayo 2014 study said that 70% of patients relapsed at some point, but it enthusiastically added that they “responded well to re-treatment with MTX”. While the 2016 Brigham and Women’s study made a similar finding, their report was more cautious and concluded with, quote, “Further investigation is needed to determine an appropriate treatment algorithm for patients with EF.”.



Last known state after 1-7 years (forums)

Forums 2017 (n=242)	
Improved	39%
Remission	28%
Relapsed	17%
Unknown	17%

Mayo 2014 (n=16)	
Improved	44%
Remission	19%
Relapsed	38%
Unknown	0%

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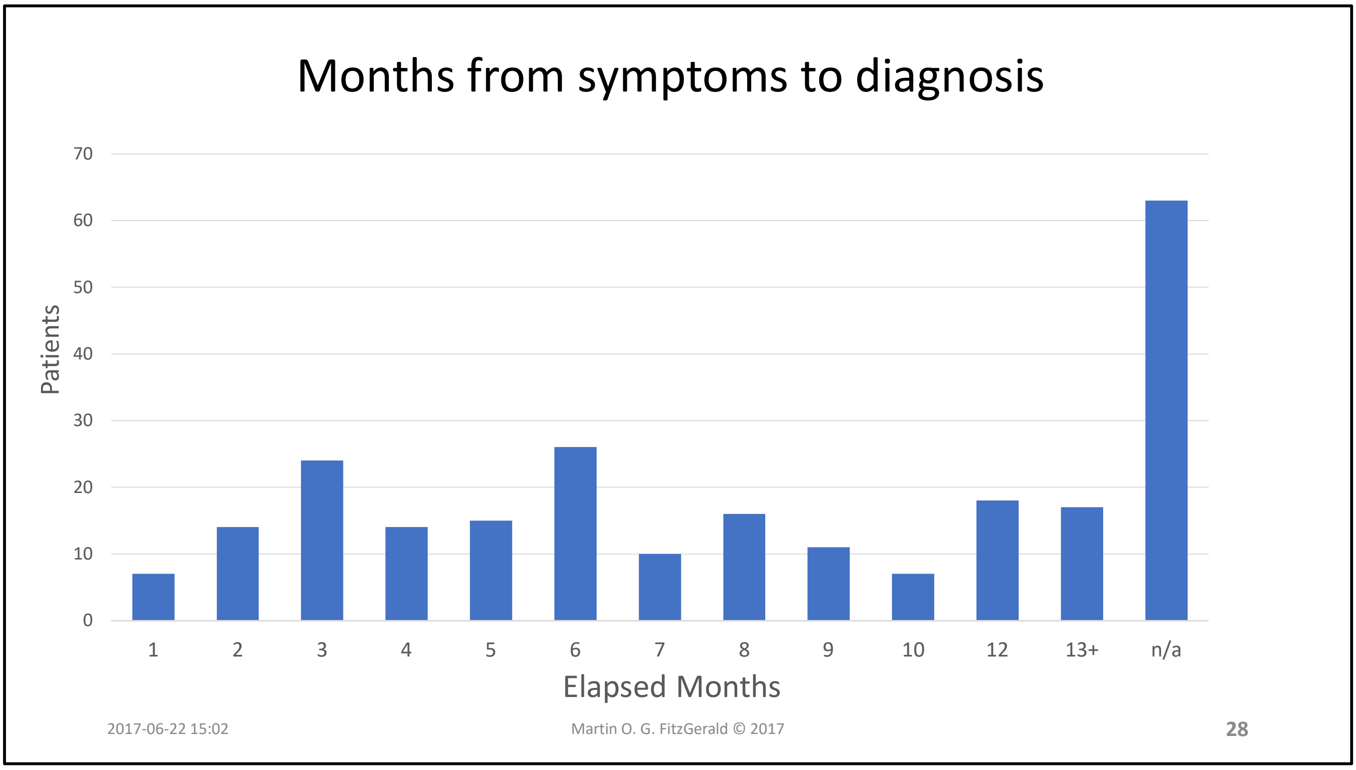
New patients ask how long (in weeks or months) until they will be “cured”. The reality is less optimistic, these are years, but it does show that most of us recover sooner or later. There are a few not shown whose history went longer than 7 years. Some went 20 years or more.



Months from symptoms to diagnosis

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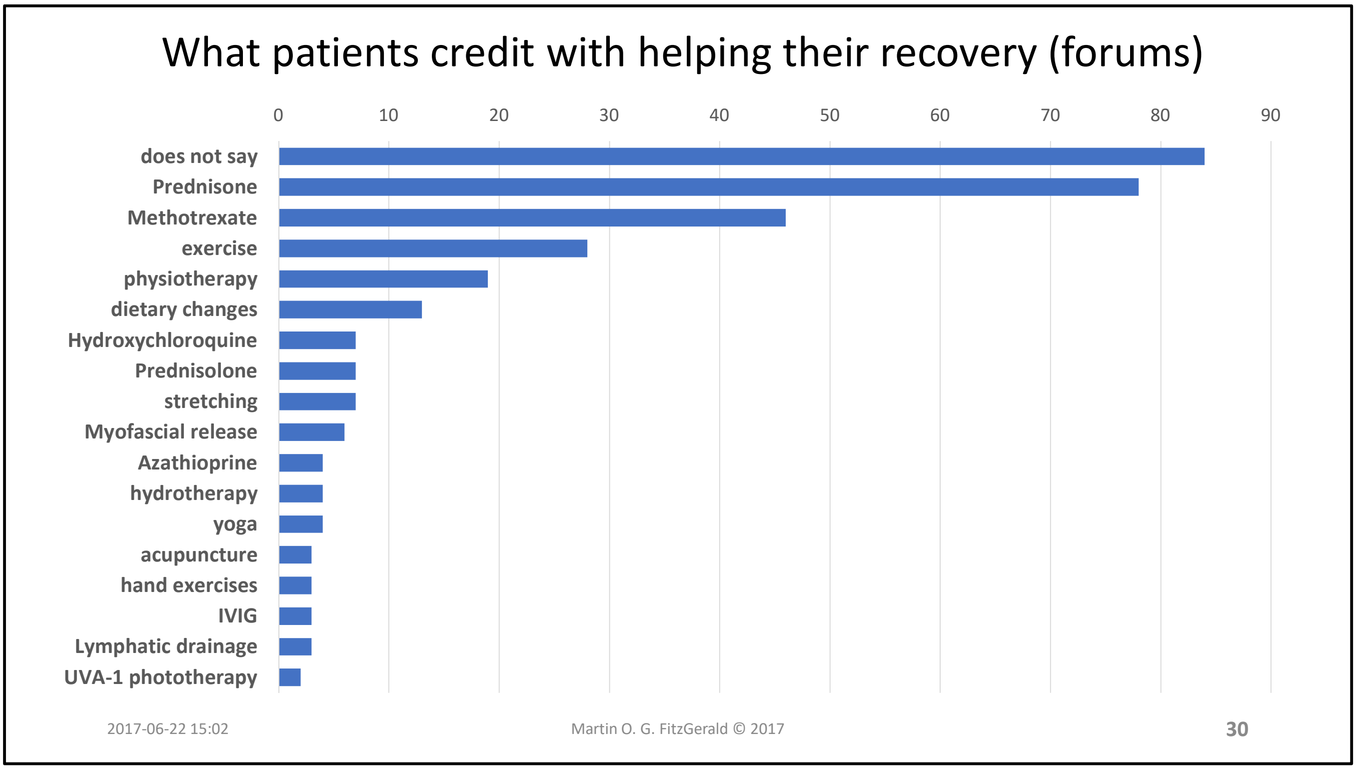
But this shows it is taking too long to diagnose this disease. Some patients lost time by delaying their first doctor visit. Quite a few complained that their doctor did not listen to them. Others were given medication on a speculative basis without a proper diagnosis. The best doctors, like my generalist, enlisted the help of others until the disorder could be identified. A very few patients were lucky enough to have a doctor who was fairly sure they knew what they had straight away, because they had already seen this disorder and recognized the symptoms.

What helped patients the most?

What helped patients the most?



What patients credit with helping their recovery (forums)

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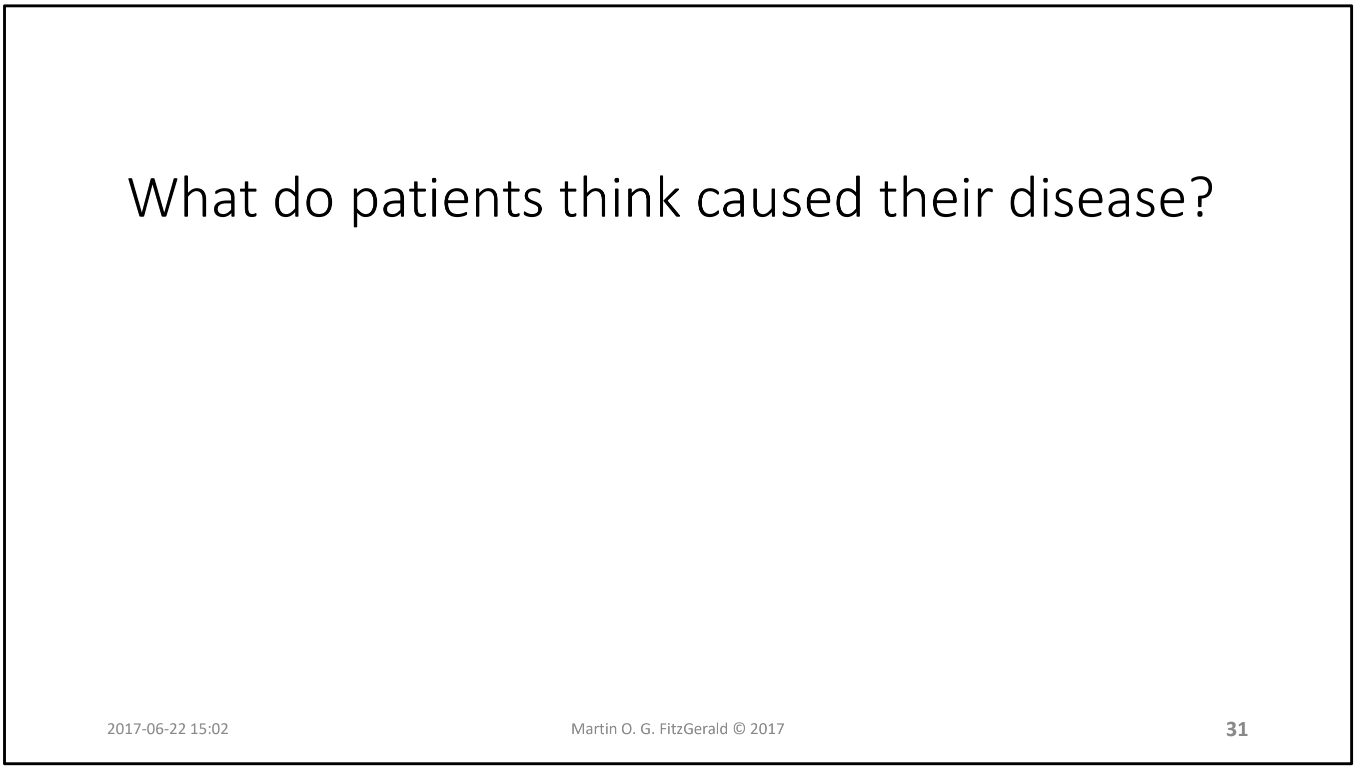
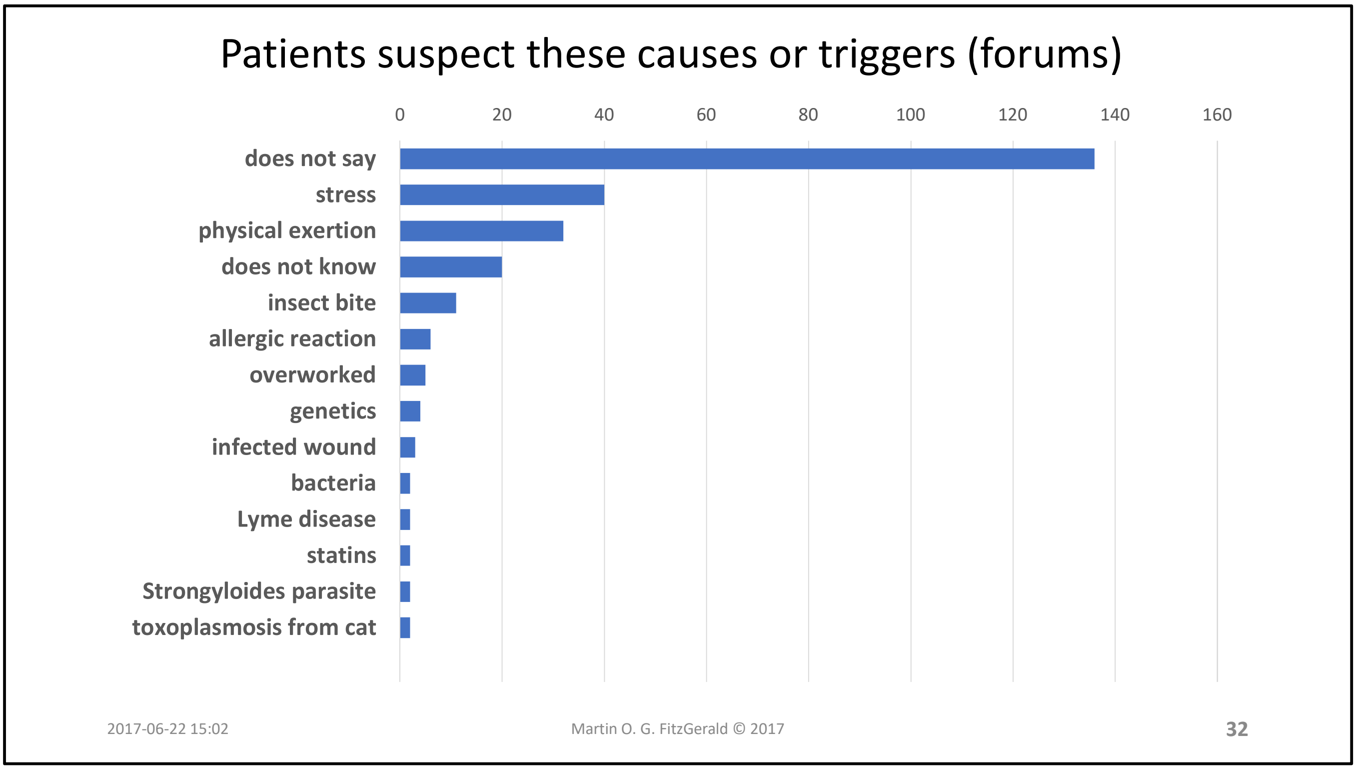
Back in 1988 the Mayo Clinic study found that about an equal number of EF patients recovered either on prednisone alone, or on hydroxychloroquine alone. In the forums very few patients were given one drug alone. Most received several so it was hard for them to know what worked best. Exercise, physical therapies and diet are given more credit by patients than by the clinical studies.



What do patients think caused their disease?

No one seems to know what causes or triggers this disease but some patients have their ideas.



Patients suspect these causes or triggers (forums)

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Stress is most often suspected by patients. Especially if we add physical “trauma” such as wounds or operations, not included here. If there was unusual physical exertion they often qualify it by remarks such as, “I have been doing this exercise for years so why should it cause a problem now?” But the stress factor is blamed without any qualifiers. Forum patients are saying it quite loudly so I think they should be heard.

What else do patients have?

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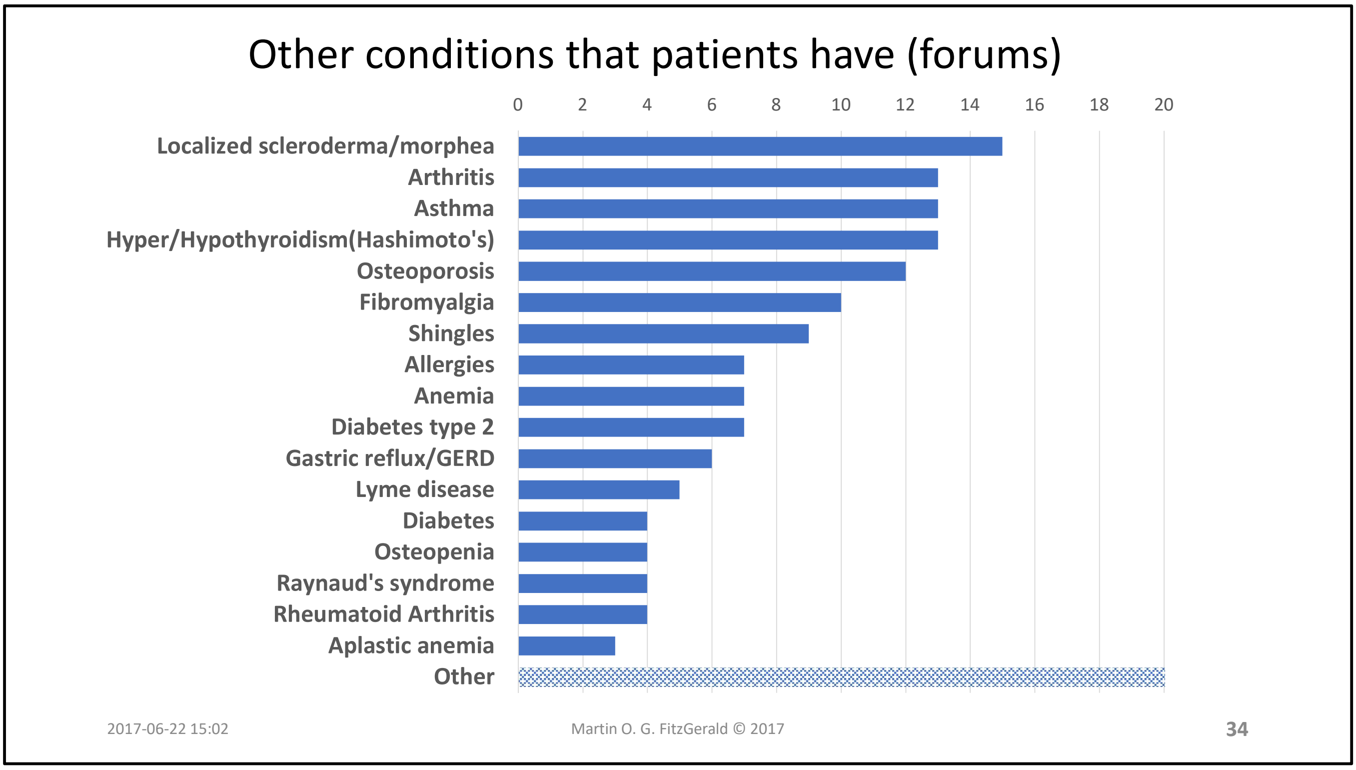

Other conditions that patients have (forums)

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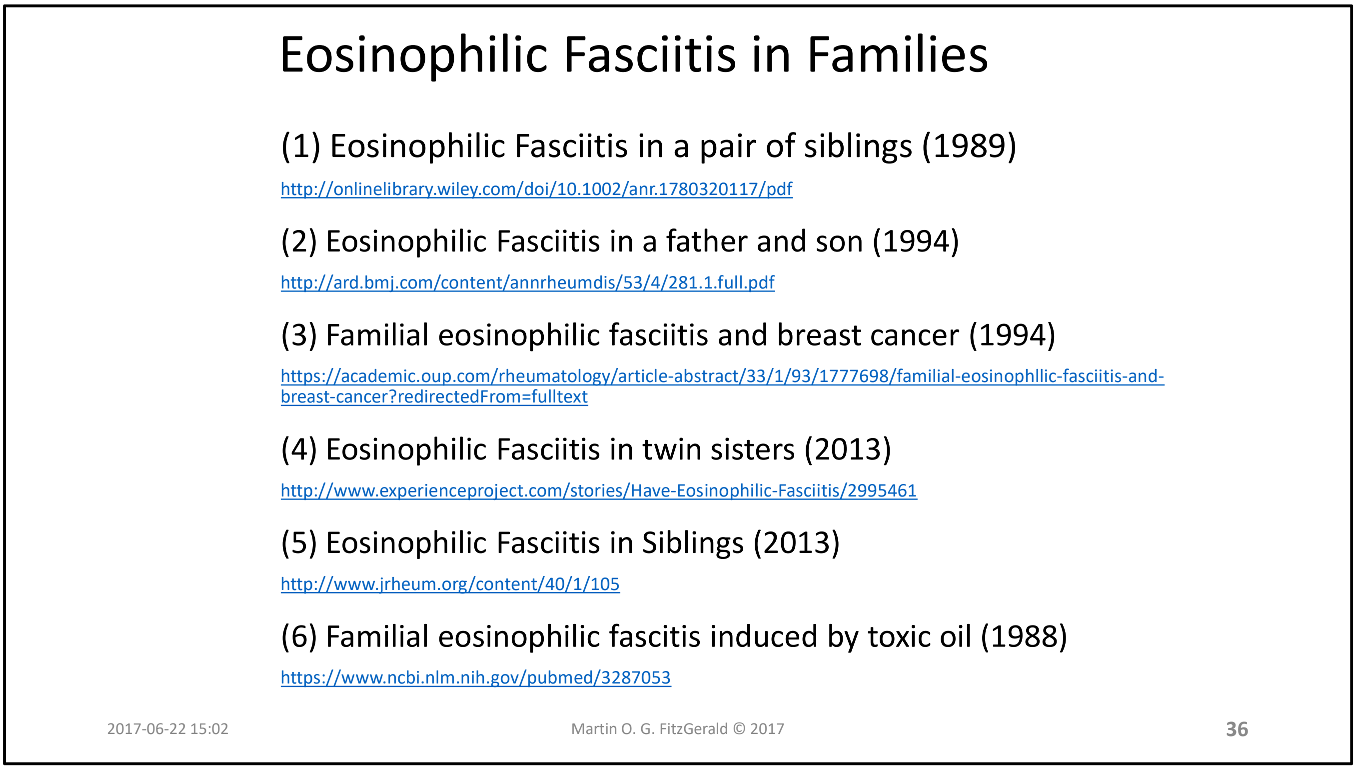
Among the 242 patients there were nearly 300 other health conditions. Some, such as Aplastic Anemia, took precedence over their EF and reduced their treatment options. “Other” goes off the screen as there are nearly 300 more. So I picked just the top ones.

Does it run in families?

Last month a woman posted that her husband, who has Eosinophilic Fasciitis, had had a related disorder, Eosinophilic Esophagitis, some years previously. Now, his son has that disorder too. This does seem very suggestive, two eosinophilic disorders in the father, one in the son.



Eosinophilic Fasciitis in Families

(1) Eosinophilic Fasciitis in a pair of siblings (1989)

http://onlinelibrary.wiley.com/doi/10.1002/anr.1780320117/pdf

(2) Eosinophilic Fasciitis in a father and son (1994)

http://ard.bmj.com/content/annrheumdis/53/4/281.1.full.pdf

(3) Familial eosinophilic fasciitis and breast cancer (1994)

https://academic.oup.com/rheumatology/article-abstract/33/1/93/1777698/familial-eosinophllic-fasciitis-and-breast-cancer?redirectedFrom=fulltext

(4) Eosinophilic Fasciitis in twin sisters (2013)

http://www.experienceproject.com/stories/Have-Eosinophilic-Fasciitis/2995461

(5) Eosinophilic Fasciitis in Siblings (2013)

http://www.jrheum.org/content/40/1/105

(6) Familial eosinophilic fascitis induced by toxic oil (1988)

https://www.ncbi.nlm.nih.gov/pubmed/3287053

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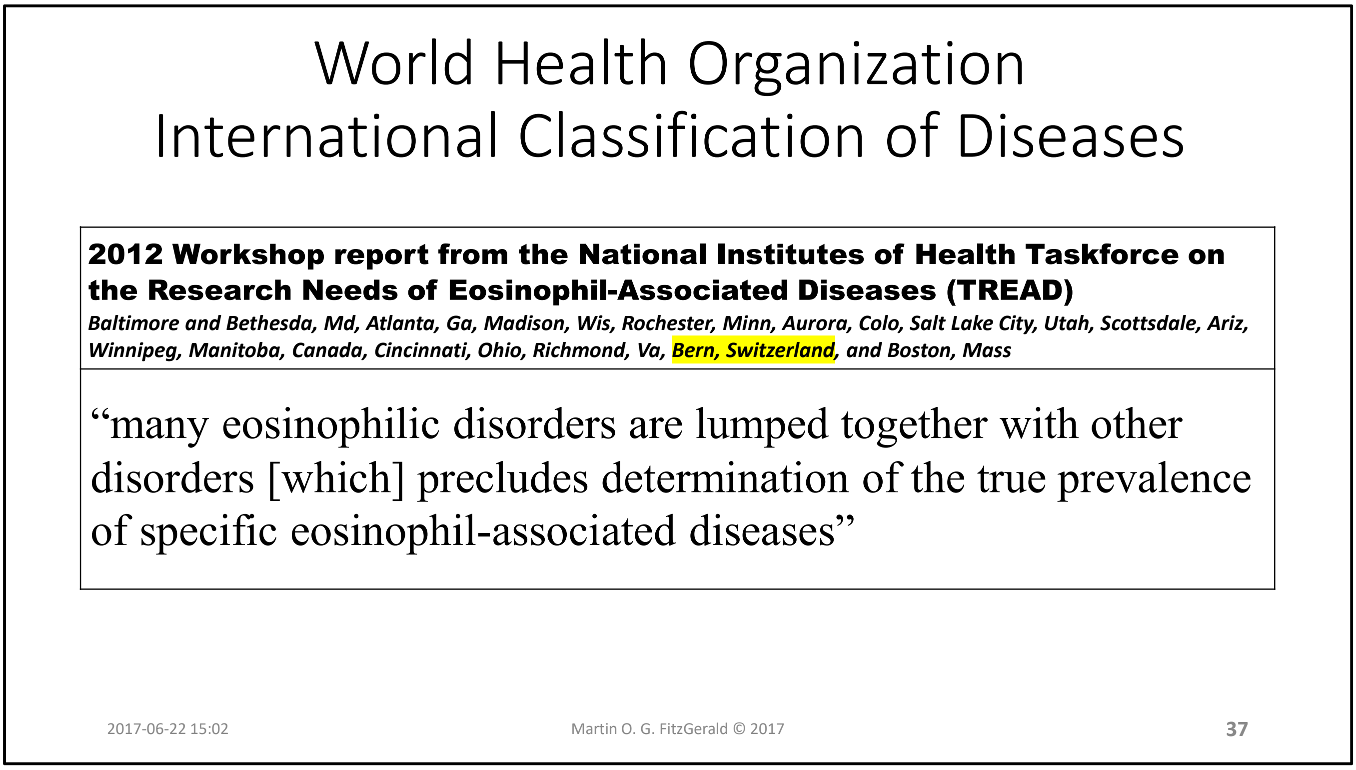
These looked to me like cases that could not be blamed on environmental factors or coincidence. I think this argues for further study into the genetic side of this disorder. The next slides look at how the W-H-O classifies eosinophilic disorders.



World Health Organization
International Classification of Diseases

2012 Workshop report from the National Institutes of Health Taskforce on the Research Needs of Eosinophil-Associated Diseases (TREAD)

Baltimore and Bethesda, Md, Atlanta, Ga, Madison, Wis, Rochester, Minn, Aurora, Colo, Salt Lake City, Utah, Scottsdale, Ariz, Winnipeg, Manitoba, Canada, Cincinnati, Ohio, Richmond, Va, Bern, Switzerland, and Boston, Mass

"many eosinophilic disorders are lumped together with other disorders [which] precludes determination of the true prevalence of specific eosinophil-associated diseases"

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I chanced upon this report. It can be summarized in that sentence taken from it, that "many eosinophilic disorders are lumped together with **other disorders** [which] precludes determination of the true prevalence of **specific** eosinophil-associated diseases". So bad news for us patients.

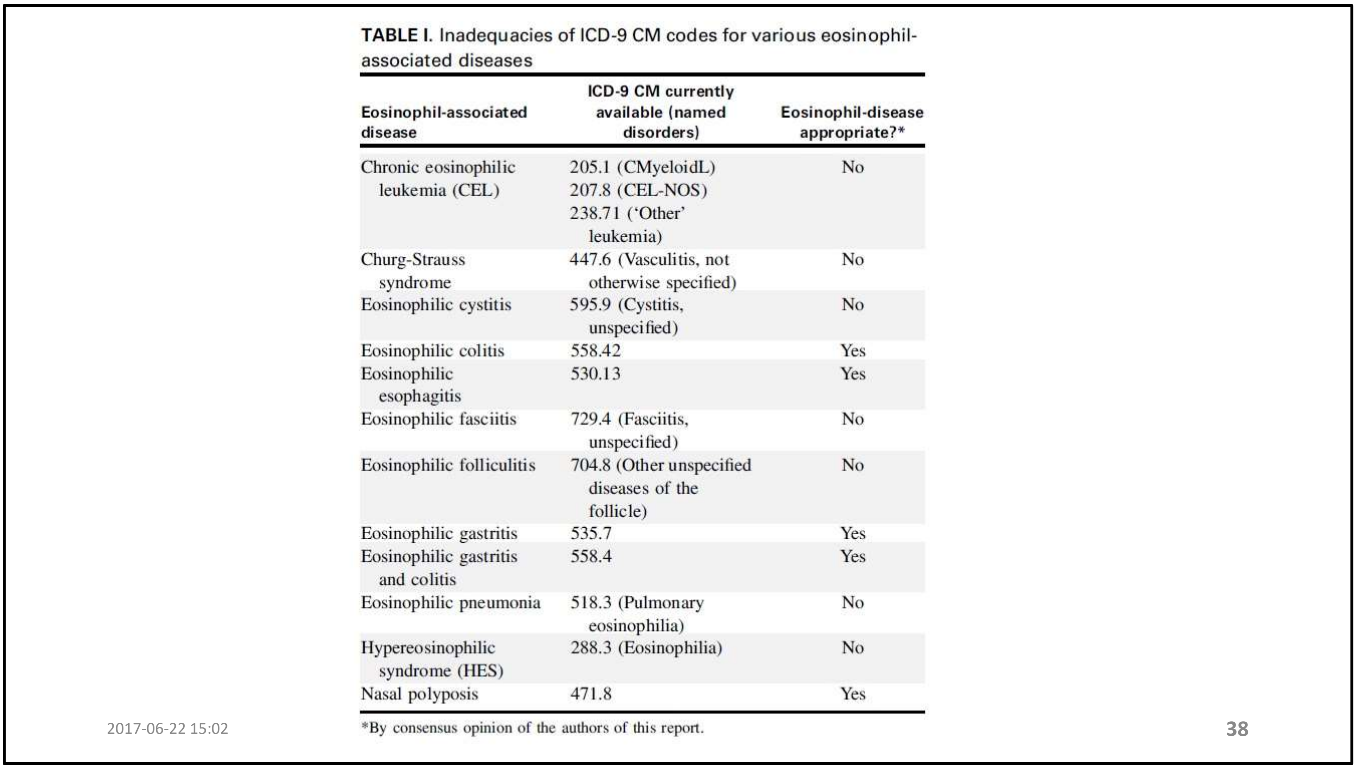
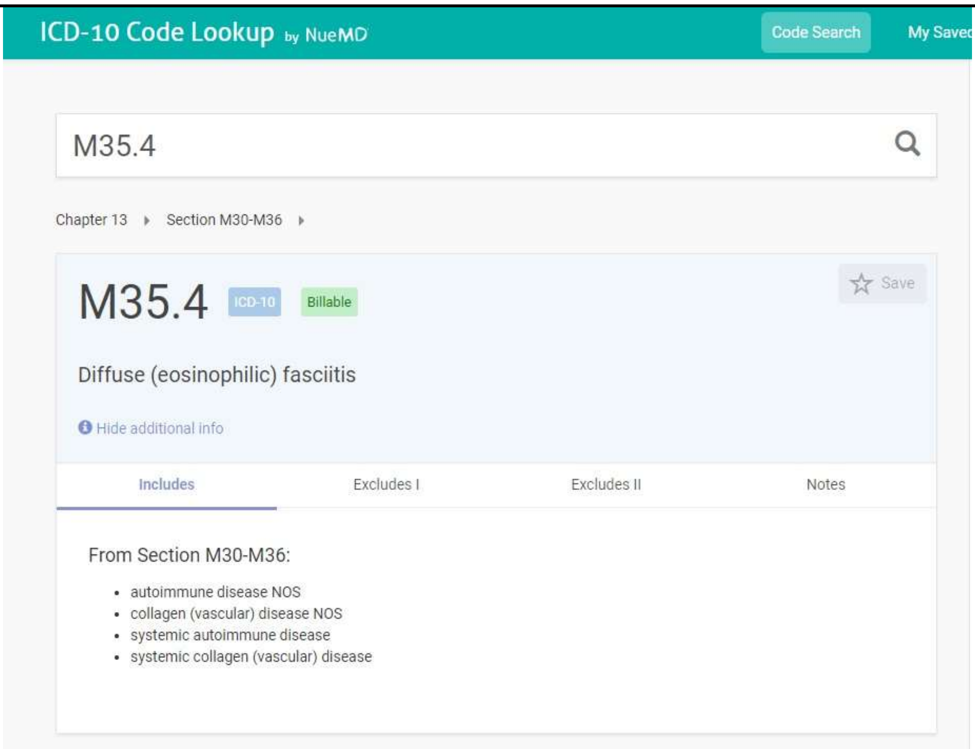


TABLE I. Inadequacies of ICD-9 CM codes for various eosinophil-associated diseases

Eosinophil-associated disease	ICD-9 CM currently available (named disorders)	Eosinophil-disease appropriate?*
Chronic eosinophilic leukemia (CEL)	205.1 (CMyeloidL) 207.8 (CEL-NOS) 238.71 ('Other' leukemia)	No
Churg-Strauss syndrome	447.6 (Vasculitis, not otherwise specified)	No
Eosinophilic cystitis	595.9 (Cystitis, unspecified)	No
Eosinophilic colitis	558.42	Yes
Eosinophilic esophagitis	530.13	Yes
Eosinophilic fasciitis	729.4 (Fasciitis, unspecified)	No
Eosinophilic folliculitis	704.8 (Other unspecified diseases of the follicle)	No
Eosinophilic gastritis	535.7	Yes
Eosinophilic gastritis and colitis	558.4	Yes
Eosinophilic pneumonia	518.3 (Pulmonary eosinophilia)	No
Hypereosinophilic syndrome (HES)	288.3 (Eosinophilia)	No
Nasal polyposis	471.8	Yes

*By consensus opinion of the authors of this report.

This table in that report shows that Eosinophilic Fasciitis is among the orphaned conditions assigned a “No” for not being appropriately classified. That was in ICD Revision 9.



M35.4

Chapter 13 ▸ Section M30-M36 ▸

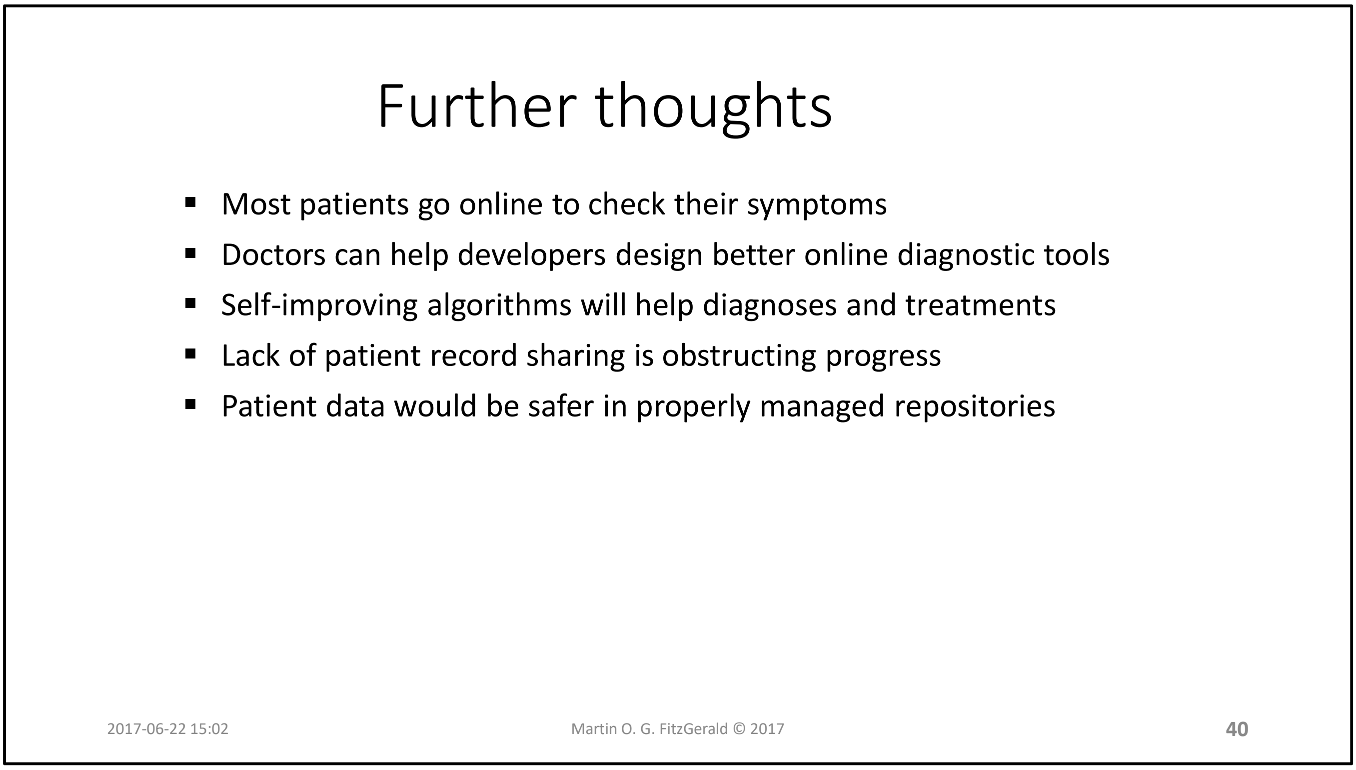
M35.4

Diffuse (eosinophilic) fasciitis

From Section M30-M36:

- autoimmune disease NOS
- collagen (vascular) disease NOS
- systemic autoimmune disease
- systemic collagen (vascular) disease

In ICD Revision 10 the disease has its own classification with a new number. According to the workshop report, quote, this should “make it easier for medical professionals and researchers”. It sounds like an opportunity for **health software** as well.



Further thoughts

- Most patients go online to check their symptoms
- Doctors can help developers design better online diagnostic tools
- Self-improving algorithms will help diagnoses and treatments
- Lack of patient record sharing is obstructing progress
- Patient data would be safer in properly managed repositories

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Just a few remarks to conclude. In a local newspaper, La Gruyère, a doctor recently warned readers that the internet was only a source of information, not a diagnostic tool. I diagnosed my whooping cough 10 years ago by listening to online recordings of coughs. My doctor was impressed as he had mis-diagnosed it as asthma.

Patients will try to self-diagnose online, so rather than resist this, health professionals could collaborate with software developers so we all benefit.

Machine learning is improving by the day, but it needs access to far more health data than what seems to be on offer at present. This will lead to earlier diagnoses and better treatments.

If you have patient data on a local computer somewhere, at the very least an offline backup will protect it from being encrypted by ransom software.

My generalist, when I told him about this talk, warned me that “most doctors are not very interested in what patients have to say”. I hope there are not too many of those in this audience.

Thank you for your time.