Israel experience with PULMONARY FIBROSIS

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Pulmonary fibrosis

- IPF has an estimated prevalence of 14-43 / 100,000
- Estimated incidence of 7-6 / 100,000
- Both prevalence and incidence increase with advancing age.

Raghu et al. Incidence and prevalence of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2006.
Fernández et al. Incidence, prevalence, and clinical course of idiopathic pulmonary fibrosis: a population-based study. Chest 2010.

Data regarding pulmonary fibrosis in north Israel

- **There is no established data regarding IPF prevalence in Israel.**

- **Carmel Medical Center and the Health Services of Haifa and the West Galil** provide medical services to 700,000 Israeli subjects.
- We revised all patients with ILD, in our clinic since 2012.

- Data of 240 patients with Intersititał lung disease was reviewed.

Data regarding pulmonary fibrosis in north Israel

- 135 patients diagnosed with pulmonary fibrosis were registered in our center, between 2012 and 2016.

- prevalence of 19.2 / 100,000.

Table 1—*Baseline Characteristics of Incident Cases Stratified by Calendar-Year of IPF Diagnosis*

Variable	Overall, 1997-2005, N = 47	Calendar-Year of IPF Diagnosis: 1997-1999, n = 20	2000-2002, n = 16	2003-2005, n = 11
Age, y				
Mean and SD	73.5 ± 7.8	74.6 ± 8.9	72 ± 7.2	74.2 ± 7.0
50-59	2 (4)	1 (5)	1 (6)	0
60-69	15 (32)	6 (30)	5 (31)	4 (36)
70-79	20 (42)	8 (40)	8 (50)	4 (36)
> 80	10 (22)	5 (25)	2 (12)	3 (27)
Men	28 (59)	11 (55)	11 (69)	6 (54)
BMI, kg/m²	27.1 ± 4.9	26.1 ± 3.8	29.2 ± 6	26.2 ± 2
Surgical lung biopsy examination-proven UIP	14 (29)	5 (25)	5 (31)	4 (36)
CT fibrosing score, %	29.8 ± 14	29.2 ± 14	30 ± 10	31 ± 18
Definite UIP on CT pattern	11 (23)	6 (30)	3 (19)	2 (18)
Nonbiopsy cases with definite UIP CT pattern	10 (21)	6 (30)	2 (12)	2 (18)
Smoking, pack-years				
< 20	5 (11)	1 (5)	3 (19)	1 (9)
20-40	14 (29)	5 (25)	6 (37)	3 (27)
> 40	9 (19)	3 (15)	3 (19)	3 (27)
Never	19 (40)	11 (55)	4 (27)	4 (36)
Pulmonary function tests, % predicted				
TLC	72.1 ± 15	68 ± 13.1	75.2 ± 12.8	74.3 ± 20
FVC	68.8 ± 18.8	65.2 ± 17.3	68.1 ± 20.2	74.2 ± 19.4
FEV_1	72.5 ± 17.7	69 ± 17.7	73.1 ± 20.5	77.3 ± 13.2
FEV_1/FVC	82.5 ± 8.5	83.2 ± 9.4	82.3 ± 8.8	82.1 ± 7.3
DLCO	49.1 ± 16.4	49.8 ± 14.9	49.8 ± 19.3	47.1 ± 15.5
SpO_2, rest	94.5 ± 1.8	94 ± 2	95.1 ± 1.7	94.6 ± 1.5
SpO_2, exercise	88.7 ± 4.5	87 ± 4.7	90.4 ± 3.8	89 ± 4.6

Fernández Pérez ER et al. Incidence, prevalence, and clinical course of idiopathic pulmonary fibrosis: a population-based study. Chest 2010; 137:129.

Demographic data:

	Data from northern Israel	Data according to Chest 2010
Prevalence	19.2/100,000	27.9/100,000
Age (years)	73.21	74.2±7
male	79%	54%
female	21%	46%
never smoked	37%	36%
past + current smokers	63%	64%

Pulmonary function tests:

	Carmel MC data	Data according to CHEST 2010
PFT:		
FVC m"l	2230	
FVC %	72.50%	74.2+/-19.2
FVC decline (m"l)	297	
DLCO %	52.4%	47.1+/-15.5
DLCO decline (%)	3.75%	
Saturation - rest	90.50%	94.6 +/-1.5
oxygen treatment	31%	

Diagnosis made by:

- **Nine** patients were diagnosed by pathological UIP pattern per **TBB cryo biopsy**.
- **Five** patient were diagnosed by pathological UIP pattern per **VATS biopsy**.
- **Two** patient were diagnosed by pathological UIP pattern on **open lung biopsy**.

- Diagnosis was made **solely by CT-** in **119** patients.

Medical treatment Indication for IPF

- For patients with mild or moderate IPF, based on pulmonary function tests (FVC>50% and DLCO> 30%),
- who do not have underlying liver disease:

➔ it is recommended to initiate anti fibrotic therapy.

Medical treatment.

- **84 patients**, with pulmonary fibrosis, started treatment with Nintedanib (Ofev) or Pirfenidine (Esbriet) .

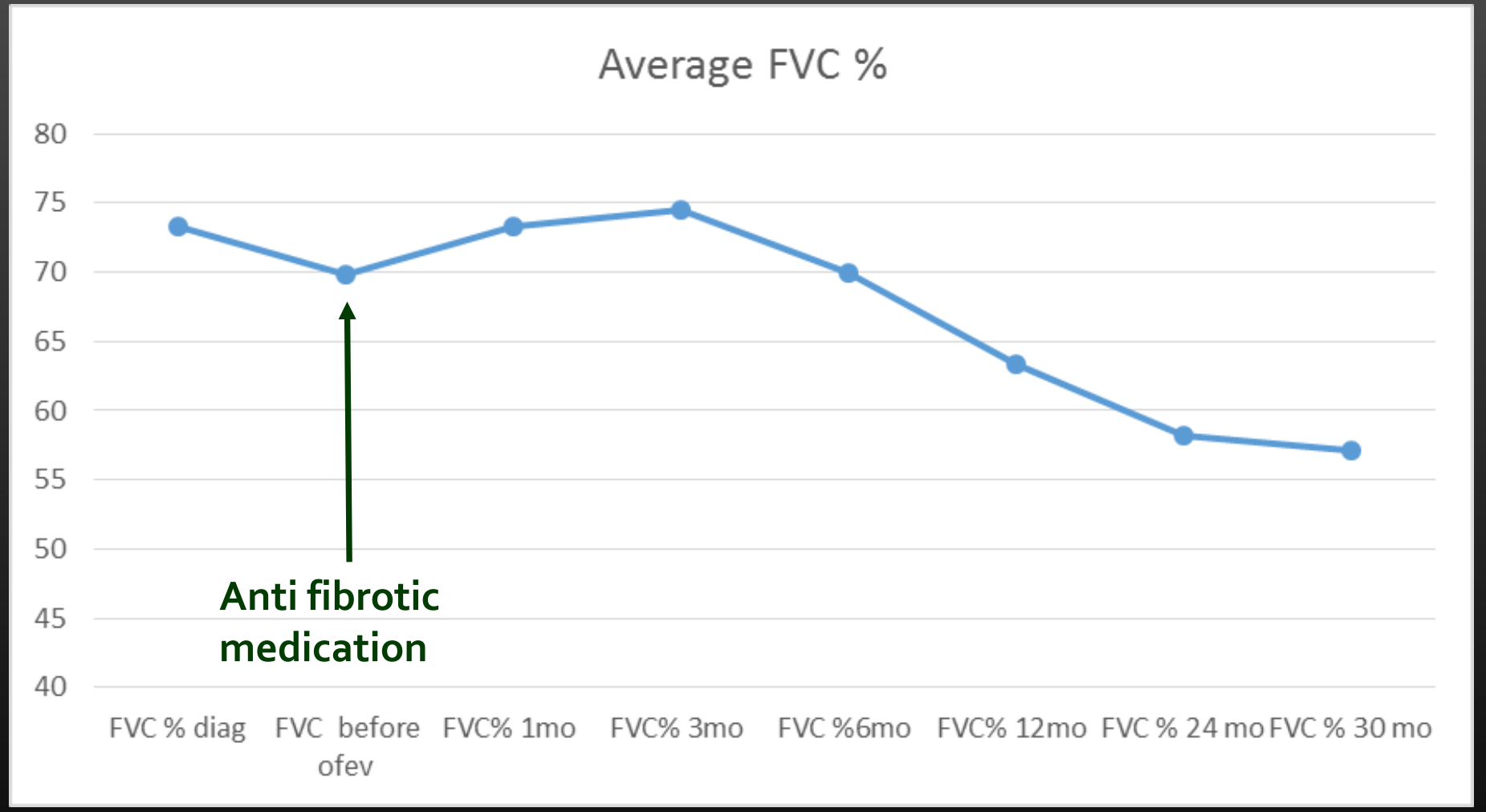
Tolerability to therapy

- 68 patients continue the treatment.
- 16 patients discontinued the medication.

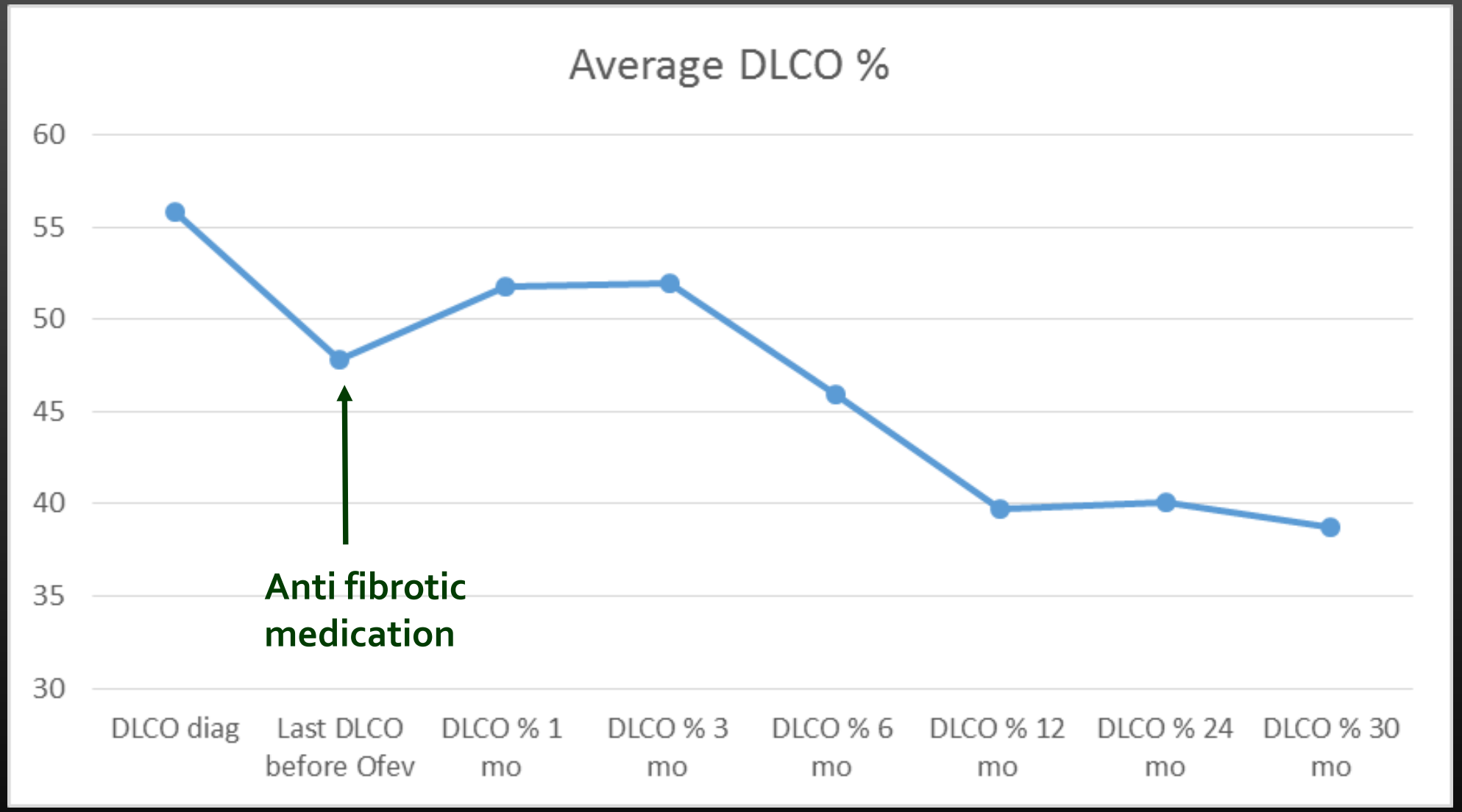
Side effects

- Frequent side effects:
- **GI**: abdominal pain, epigastric pain, GERD, nausea, vomiting, weight loss, constipation, LFT elevation.
- With Pirfenidone –skin rash and photosensetivity also seen.
- **Other**: fever, fall, weakness, hyperkalemia, epistaxis, face edema, viral infection.

- One patient had an acute coronary syndrome and one had transient ischemic attack while on Nintedanib, (The patients were previously known to have IHD and PVD respectively). Relationship of events to medication use was not certain.



Average Forced Vital capacity during antifibrotic therapy



Average diffusing lung capacity, during antifibrotic therapy

Conclusions

- With anti fibrotic therapy, some patients have side effects, mostly abdominal pain, vomiting and diarrhea.
- More severe side effects: ACS, TIA, elevated liver enzymes, weight loss.
- Most patients continued the treatment (some with dose reduction).
- Close monitoring is advised.

- Outcome- the rate of pulmonary function deterioration may be reduced.

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