DIFFERENT ILD CASES: Case Discussion

Dr. Gamze KIRKIL

Firat University

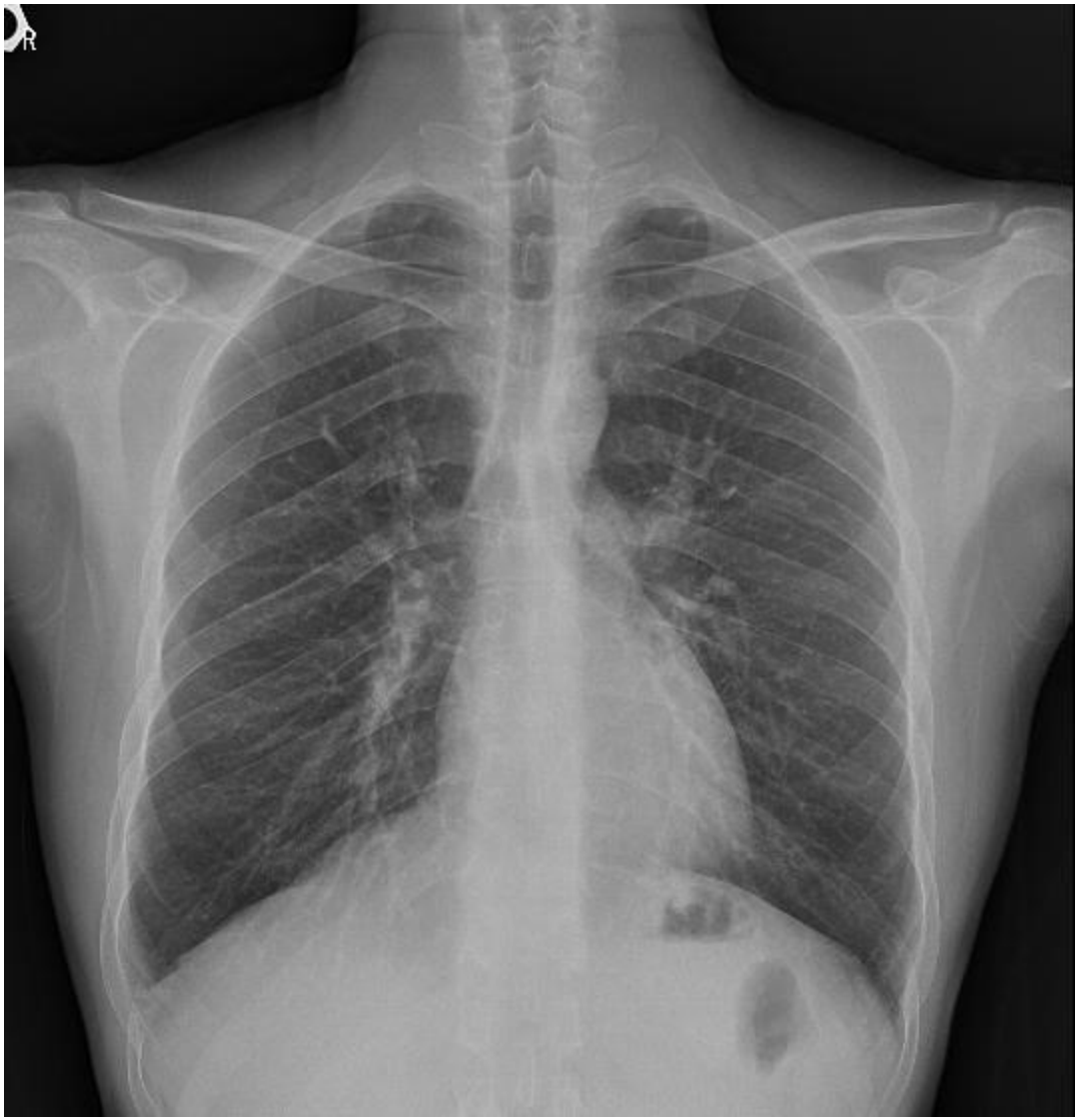
Chest Disease Department

Case-1

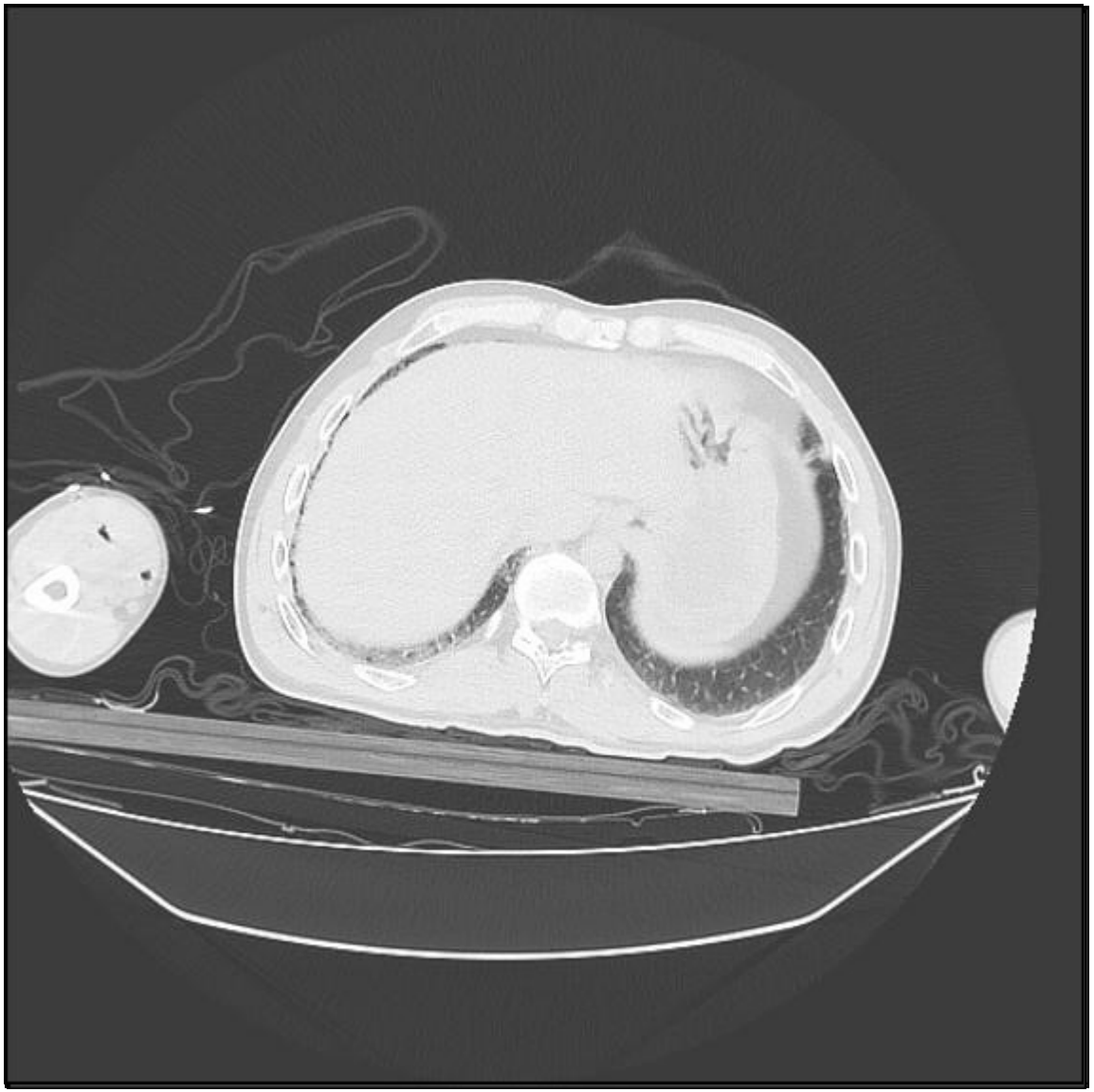
- 39 y, M
- Symptoms: Dyspnea, dry cough
- Smoking: 20 pack/y, ex-smoker for 2 years
- Occupation: Accountant
- Contact with parrot at home for years
- Medication: Lyrica cap
- Family history: Mother breast Ca, father prostate Ca

- He referred to our outpatient clinic after detecting pathology in the lungs on abdominal CT taken after gunshot in 2021
- No dyspnea, no cough
- PE: Good apperance, TA: 120/80 mmHg, Fever: 36.7 C, Pulse: 75/min, SpO2: %98
- No pathology on respiratory system exam

10.02.2021



22.02.2021



T.C.
FIRAT ÜNİVERSİTESİ HASTANESİ
RADYOLOJİ RAPORU

Adı Soyadı	:	**Rapor Tarihi**	: 26.02.2021 10:38
T.C Kimlik No	:	**Dosya no**	: 1054964
Baba Adı	: NİHAT	**Başvuru No**	: 9879666
Kurumu	: SSK SAĞLIK İŞLERİ MÜDÜRLÜĞÜ	**Doğum Yeri - Tarih**	: ELAZIĞ - 1985 **Yaş:** 35
İstem Tarihi	: 22.02.2021(20984925)	**İstem Kabul Tarihi**	: 22.02.2021(46261)
Hizmet Adi	: BT, TORAKS	**Cinsiyet**	: E

Tanı :	**Kodu**	**Adı**
	E55.9	VİTAMİN D EKSİKLİĞİ, TANIMLANMAMIŞ
	K21	GASTRO-ÖZOFAJİAL REFLÜ HASTALIĞI
	K58	İRRİTABL BARSAK SENDROMU

ÇOK KES?TL? B?LG?SAYARLI TOMOGRAF?
TORAKS

Teknik : 70 ml kontrast madde verilerek yap?lan çok kesitli BT tetki?inde;

Brakiosefalik vasküler yap?lar, trake ve ana bron?lar, özefagus normal görünümdedir.
Kalp ve ana vasküler yap?lar normal boyutlarda olup patoloji izlenmemi?tir.
Bilateral akci?er üst lob apikallerde plevral kal?nla?malar?n e?lik etti?i parankimal fibrotik de?i?iklikler izlenmektedir.
Bilateral akci?erlerde büllöz amfizematöz de?i?iklikler izlendi.
bilateral akci?er periferinde fibrotik de?i?iklikler buzlu cam görünümleri ve bal pete?i görünümleri izlendi (?nterstisiyel akci?er hastal???).
Sa? humerus ba?? ve sa? skapulada milimetrik alan izlendi (Kemik adac??? ?).
Gö?üs duvar?, kemik yap?lar ve yumu?ak dokular normal görünümdedir.

Thorax CT: Bilaterally peripheral fibrotic changes, ground-glass opacity, honeycomb

Rheumatologic biomarkers (-)

Dosya No	1054964			BUL
Başvuru No				
Sonuç Durumu	☑ Bekleyenler	☑ Onaylanacaklar	☑ Tamamlanmışlar	
☐ Referans Aralığı Kontrol				

Başvuru Tarihi	Başvuru No	Alt Birim Adı
		Tüm Başvurular
24.09.2024 08:57	14434030	GÖĞÜS HASTALIKLARI POLİKLİNİ...
02.07.2024 09:32	14136810	GÖĞÜS HASTALIKLARI POLİKLİNİ...
12.02.2024 10:15	13645577	GÖĞÜS HASTALIKLARI POLİKLİNİ...

Barkod	Kabul Tarihi	N.K	N.K. Tarih	Test Adı	RF	Parametre ...		Sonuç	Birim	T.Sonuç	Durum	Alt Limit
00011128109	10.03.2021 10:50	+	10.03.2021 11:45	ANA 1/100 SERUM DİL...		ANA(HEP2) ...		Negatif				
00011096738	02.03.2021 09:47	+	02.03.2021 12:19	ANTİ ENDOMİSYUM 1/...		ANTİ ENDO...		Negatif				
00011128108	10.03.2021 10:50	+	10.03.2021 11:34	CRP	▼	CRP		<3.3	mg/L			0
00011128108	10.03.2021 10:50	+	10.03.2021 11:34	RF	▼	RF		<9.38	IU/mL			0
00011128106	10.03.2021 10:50	+	10.03.2021 11:36	CCP		CCP		< 0.5	u/mL			0

Barkod	Kabul Tarihi	N.K	N.K. Tarih	Test Adı	RF	Parametre ...		Sonuç	Birim	T.Sonuç	Durum	Alt Limit
00011128106	10.03.2021 10:50	+	10.03.2021 11:35	ANTİ-SCL 70		ANTİ-SCL 70		4.70	IU/ml		Negatif	0
00011128104	10.03.2021 10:50	+	10.03.2021 11:36	ANTİ SM	▼	ANTİ SM		<3.0	IU/ml		Negatif	0
00011128104	10.03.2021 10:50	+	10.03.2021 11:36	ANTİ-DS DNA (ELISA)		ANTİ-DS DN...		<10.0	IU/ml		Negatif	0
00011128104	10.03.2021 10:50	+	10.03.2021 11:36	ANTİ U1 RNP		ANTİ U1 RNP		12.40	IU/ml			0
00011128104	10.03.2021 10:50	+	10.03.2021 11:36	ANTİ-SSB LA	▼	ANTİ-SSB LA		<3.0	IU/ml		Negatif	0
00011128104	10.03.2021 10:50	+	10.03.2021 11:36	ANTİ SENTROMER (ELI...	▼	ANTİ SENTR...		<3.0	IU/ml		Negatif	0
00011096737	02.03.2021 09:47	+	02.03.2021 11:00	DOKU TRANSGLUTAMI...	▼	DOKU TRAN...		<3.0	U/ml		Negatif	0

PFT: FEV1: %75, FVC: %84, FEV1/FVC: %82

T.C
FIRAT ÜNİVERSİTESİ
PATOLOJİ LABARATUVARI
PATOLOJİ RAPORU

BİYOPSİ RAPORU

Hasta Adı Soyadı		Biyopsi/Sitoloji No	S-1736/21
Biyopsi/Sitoloji No	**S-1736/21**	Isteyen Bölüm	GÖĞÜS HASTALIKLARI KLİNİĞİ
T.C. Kimlik No	3734*******	Tetkiki Isteyen Doktor	MUTLU KULUÖZTÜRK .
Yaş / Cinsiyeti	36 / E	Rapor Ilk Kayıt Tarihi	
DosyaNo	1054964		

Tetkik Istem Zamanı	01.04.2021 11:27	Numune Kabul Zamanı	01.04.2021 13:58
Numune Alma Zamanı	01.04.2021 11:27	Uzman Onay Zamanı	13.04.2021 16:43
Rapor Onay Tarihi	12.04.2021 13:03	Istem Tarihi	01.04.2021(21271564
Rapor Kes. Tarihi	13.04.2021 16:43	Istem Kabul Tarihi	01.04.2021(909330)

KLİNİK ÖZET:

KLİNİK BİLGİ:

MAKROSKOB?: Kapta gönderilen 20 cc berrak renkte mayi (2PAP,2MGG)

M?KROSKOB?:
-Bron? epitel hücreleri
-Alveolar makrofajlar
-Bu k?smi metaplazi de?i?iklikler gösteren skuamöz hücreler
-Yayg?n bakteri kümeleri
-Lenfositten bask?n iltihap hücreleri

TANI:
AKC??ER, BRON? LAVAJ S?TOLOJ?S? VE HÜCRE BLO?U:
-AKT?F KRON?K ?LT?HAB? REAKS?YON

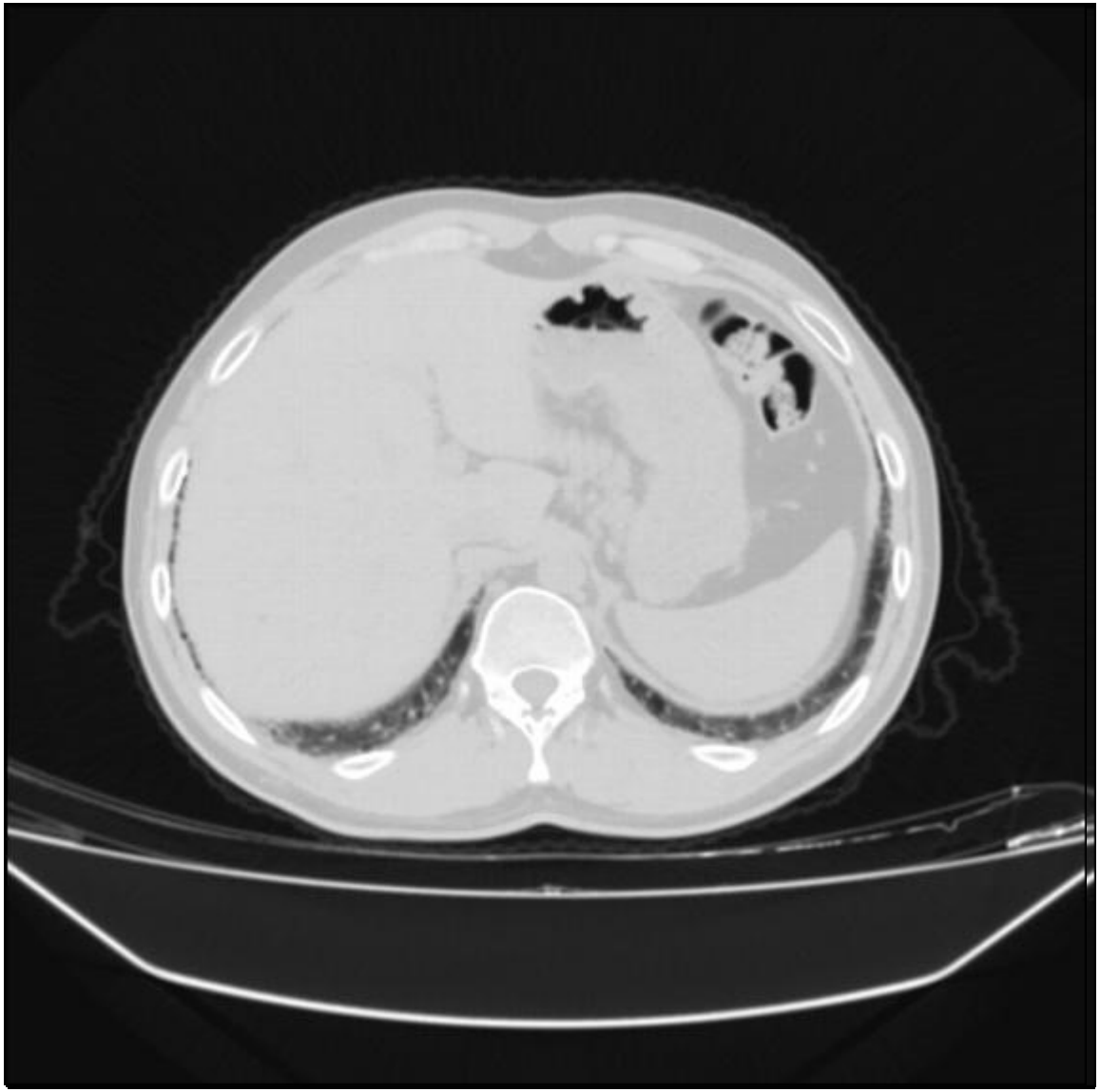
Cytology report of bronchial lavage:
Lymphocyte dominant inflammation

- Steroid is prescribed
- But he did not want to use the medicine because of side effects
- He is recommended for a control after 3 months

2th visit: October 2022

- Dyspnea and cough for 1 week
- PE: Inspiratory crackles on left medial-lower zone, right lower zone, SpO2: %96, clubbing (-), pretibial edema (-)

14.10.2022



- PFT: FEV1: %61, **FVC: %68**, FEV1/FVC: %80, **DLCO: %47**
- Pulmonary biopsy is recommended
- Cryobiopsy is applied

TIBBİ PATOLOJİ ANABİLİM DALI
PATOLOJİ RAPORU

Hasta Adı Soyadı		Biyopsi/Sitoloji No	**B-16265-2023**
T.C. Kimlik No	37348275616	İsteyen Bölüm	GOĞÜS CERRAHİ SERVİSİ
Yaş / Cinsiyeti	38 / E	Tetkiki İsteyen Doktor	
DosyaNo	37348275616		
Numune Türü	Tetkik İstem Zamanı 09.08.2023 11:28	Numune Kabul Zamanı	09.08.2023 11:30
	Numune Alma Zamanı 09.08.2023 11:28	Uzman Onay Zamanı	18.08.2023 08:55
Eski Biyopsi No			

KLİNİK BİLGİ:

AC FİBROZİS ETY..

MAKROSKOPİ:

Kayıtsız poşette gönderilen büyüğü 0,8x0,4x0,4 cm küçüğü 0,3x0,3x0,3 cm ölçüsünde toplam 6 adet bej - kahverenkli nodüler dokunun tamamı 1-2,2 kasette takibe alındı.fs/ ega

MİKROSKOPİ:

Kesitlerde, bronş akciğer parankimi çok geniş kistik lezyonlar görülmüştür. İnterstisyel mesafeler kalın, alveoller makrofajla doludur. Bazı makrofajlarda multinükleer dev hücre oluşumu gözlenmiştir.İmmunohistokimyasal olarak CD34, Pan Keratin, Vimentin, P63, TTF-1, CD68, CD163, CD1a, S-100, Desmin ve Aktin uygulanmıştır. Kistik mesafelerin büllöz amfimatöz değişim gösteren alveol alanları olduğu anlaşılmıştır. İnflmatuar reaksiyon son derece az olup, makrofajlardan oluşmakta ve alveol lümenine sınırlı kalmaktadır. Değişiklikler interstisyel akciğer hastalığı ve son dönem akciğer hastalığı - bal peteği akciğer görünümü ile uyumludur.

Biopsy report: Interstitial space fullfilled with alveolar macrophages, honeycomb apperance

- Nintedanib prescribed for **IPF** diagnosis
- He did not want to use nintedanib because of drug content
- He applied to our out-patient clinic

American Journal of Respiratory and Critical Care Medicine Volume 205 Number 9 | May 1 2022

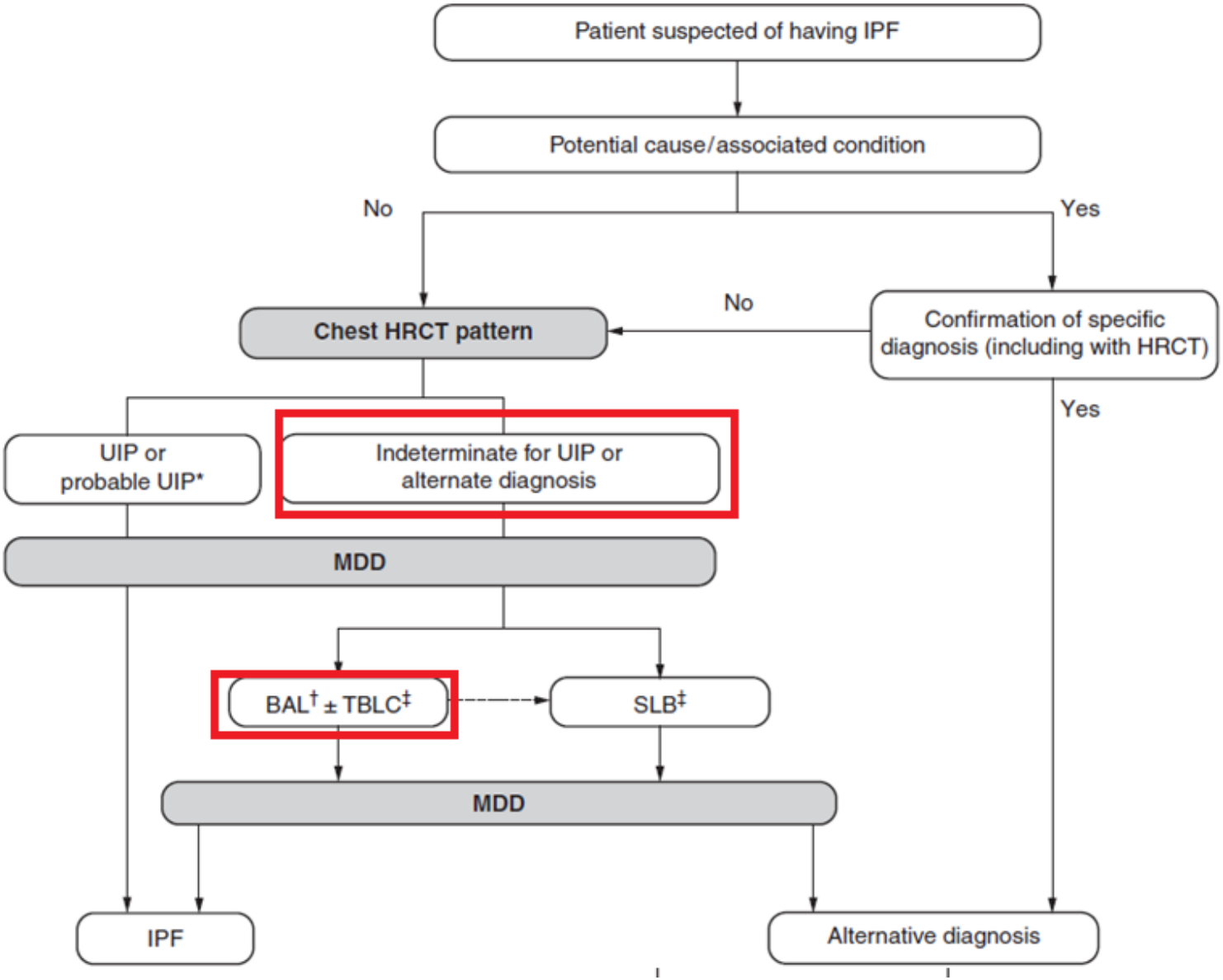
Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Table 3. High-Resolution Computed Tomography Patterns in Idiopathic Pulmonary Fibrosis

	HRCT Pattern			
	UIP Pattern	**Probable UIP Pattern**	**Indeterminate for UIP**	**CT Findings Suggestive of an Alternative Diagnosis**
Level of confidence for UIP histology	Confident (>90%)	Provisional high confidence (70–89%)	Provisional low confidence (51–69%)	Low to very low confidence (≤50%)
Distribution	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with fibrosis) • Occasionally diffuse • May be asymmetric	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with reticulation and traction bronchiectasis/ bronchiolectasis)	• Diffuse distribution without subpleural predominance	• Peribronchovascular predominant with subpleural sparing (consider NSIP) • Perilymphatic distribution (consider sarcoidosis) • Upper or mid lung (consider fibrotic HP, CTD-ILD, and sarcoidosis) • Subpleural sparing (consider NSIP or smoking-related IP)
CT features	• Honeycombing with or without traction bronchiectasis/ bronchiolectasis • Presence of irregular thickening of interlobular septa • Usually superimposed with a reticular pattern, mild GGO • May have pulmonary ossification	• Reticular pattern with traction bronchiectasis/ bronchiolectasis • May have mild GGO • Absence of subpleural sparing	• CT features of lung fibrosis that do not suggest any specific etiology	• Lung findings ◦ Cysts (consider LAM, PLCH, LIP, and DIP) ◦ Mosaic attenuation or three-density sign (consider HP) ◦ Predominant GGO (consider HP, smoking-related disease, drug toxicity, and acute exacerbation of fibrosis) ◦ Profuse centrilobular micronodules (consider HP or smoking-related disease) ◦ Nodules (consider sarcoidosis) ◦ Consolidation (consider organizing pneumonia, etc.) • Mediastinal findings ◦ Pleural plaques (consider asbestosis) ◦ Dilated esophagus (consider CTD)

etc.)
• Mediastinal findings
◦ Pleural plaques (consider asbestosis)
◦ Dilated esophagus (consider CTD)


Patient suspected of having IPF
Potential cause/associated condition
No
Yes
Chest HRCT pattern
No
Confirmation of specific diagnosis (including with HRCT)
Yes
UIP or probable UIP*
Indeterminate for UIP or alternate diagnosis
MDD
BAL† ± TBLC‡
SLB‡
MDD
IPF
Alternative diagnosis

AMERICAN THORACIC SOCIETY DOCUMENTS

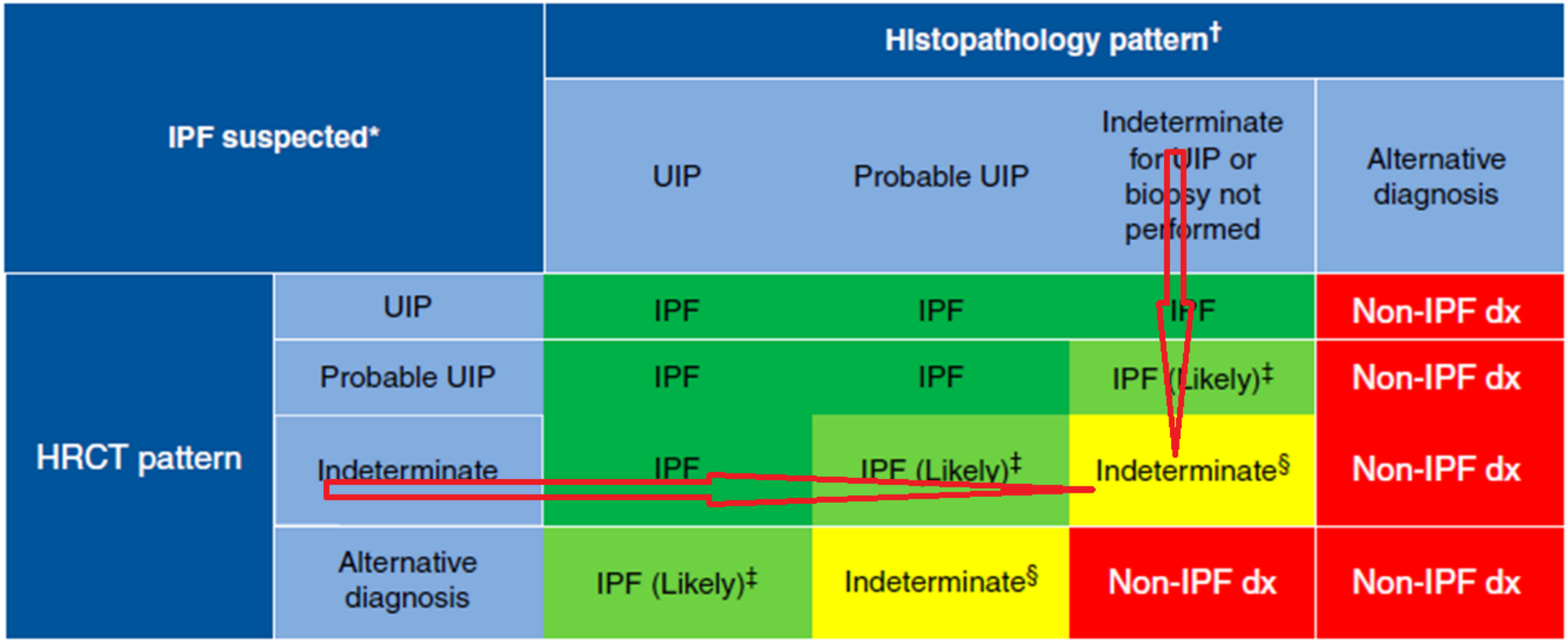
Diagnosis of Idiopathic Pulmonary Fibrosis

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

This official ATS/ERS/JRS/ALAT Clinical Practice Guideline was endorsed by the Pulmonary Pathology Society October 2018

Table 5. Histopathology Patterns and Features

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
• Dense fibrosis with architectural distortion (i.e., destructive scarring and/or honeycombing) • Predominant subpleural and/or paraseptal distribution of fibrosis • Patchy involvement of lung parenchyma by fibrosis • Fibroblast foci • Absence of features to suggest an alternate diagnosis	• Some histologic features from column 1 are present but to an extent that precludes a definite diagnosis of UIP/IPF *And* • Absence of features to suggest an alternative diagnosis *Or* • Honeycombing only	• Fibrosis with or without architectural distortion, with features favoring either a pattern other than UIP or features favoring UIP secondary to another cause* • Some histologic features from column 1, but with other features suggesting an alternative diagnosis†	• Features of other histologic patterns of IIPs (e.g., absence of fibroblast foci or loose fibrosis) in all biopsies • Histologic findings indicative of other diseases (e.g., hypersensitivity pneumonitis, Langerhans cell histiocytosis, sarcoidosis, LAM)


IPF suspected*
Histopathology pattern†
UIP
Probable UIP
Indeterminate for UIP or biopsy not performed
Alternative diagnosis
HRCT pattern
UIP
IPF
IPF
IPF
Non-IPF dx
Probable UIP
IPF
IPF
IPF (Likely)‡
Non-IPF dx
Indeterminate
IPF
IPF (Likely)‡
Indeterminate§
Non-IPF dx
Alternative diagnosis
IPF (Likely)‡
Indeterminate§
Non-IPF dx
Non-IPF dx

Re-evaluation of biopsy sample in a different hospital

Gönderilen Materyal : KONSÜLTASYON Materyallerin Alındığı Yer : AKCİĞER Materyalin Alınma Şekli : 5 Klinik Ön Tanı : İntersitisyel akciğer hastalığı son dönem bulguları ile uyumlu değişiklikler Makroskopi : Konsültasyon amacıyla gönderilen B- 16265-2023 nolu 14 adet lam+ 2 adet blok tarafımıza gönderilmiştir. Döküm, Kesit Tek: Simay Gök, Rapor Sek: Öznur Bozkurt Mikroskopi : Histokimyasal Boyama Panel Sonuçları : MTK uygulandı İmmunhistokimya Boyama Panel Sonuçları : Frozen Tanı : Histopatalojik Tanılar / Sitopatolojik Tanılar : Akciğer, Eksizyonel Biopsi; Konsültasyon: A- Tip II pnömosit proliferasyonu, alveol septaları kalınlaştıran lenfoid proliferasyon, alveol boşluklarında makrofajlar, fibrozis, düz kas metaplazi odağı B- Tip II pnömosit proliferasyonu, alveol septaları kalınlaştıran fibrozis,	01.09.2023	T.C. SAĞLIK BAKANLIĞI İSTANBUL YEDİKULE GÖĞÜS HASTALIKLARI VE GÖĞÜS CERRAHİSİ EAH	GÖĞÜS HASTALIKLARI	08.09.2023

Report: Type II pneumocyte proliferation, lymphocyte aggregation thickens alveolar septa, macrophages in alveolar sac, fibrosis, smooth muscle metaplasia focus

İSTANBUL VALİLİĞİ
İL SAĞLIK MÜDÜRLÜĞÜ
S.B.Ü. İstanbul Yedikule Göğüs Hastalıkları Ve Göğüs Cerrahisi Eğitim Ve Araştırma Hastanesi

PATOLOJİ RAPORU

Bas Tar	11.09.2023
Protokol Kayıt No	2023 / 8360
Onay Tarihi	08.09.2023 19:33

Hastanın Adı Soyadı	[redacted]	**Laboratuar Kabul Tarihi\Saati**	Eylül 01 2023, 11:50
Hasta T.C. Kimlik No	[redacted]	**Patoloji Rapor Tarihi**	Eylül 08 2023, 19:33
Doğum Tarihi, Cinsiyeti	20.03.1985 / E	**Protokol No**	
İsteyen Birim	48.4 Göğüs Polikliniği D. TURAN	**Rapordan Sorumlu I.Doktor**	[redacted]
İsteyen Hekim	DEMET TURAN	**Raporlayan Sekreter**	
İstemin Yapıldığı Tarih\Saati	01.09.2023 11:03:40	**Numune Alma Tarihi ve Saati**	

Gönderilen Materyal KONSÜLTASYON

Materyallerin Alındığı Yer AKCİĞER **Materyalin Alınma Şekli :** KONSÜLTASYON

Klinik Ön Tanı

İnterstisyel akciğer hastalığı son dönem bulguları ile uyumlu değişiklikler

Makroskopi

Konsültasyon amacıyla gönderilen B- 16265-2023 nolu 14 adet lam+ 2 adet blok tarafımıza gönderilmiştir.

Döküm, Kesit Tek: Sinay Gök, Rapor Sek: Öznur Bozkurt

TIBBİ LABORATUVAR YORUMU

Bulgular fibrotik tip Nonspesifik İnterstisyel Pnömoni paterni ile uyumludur. Olgunun kollagen doku hastalıkları yönünden tetkiki önerilir

Findings are compatible with fibrotic NSIP

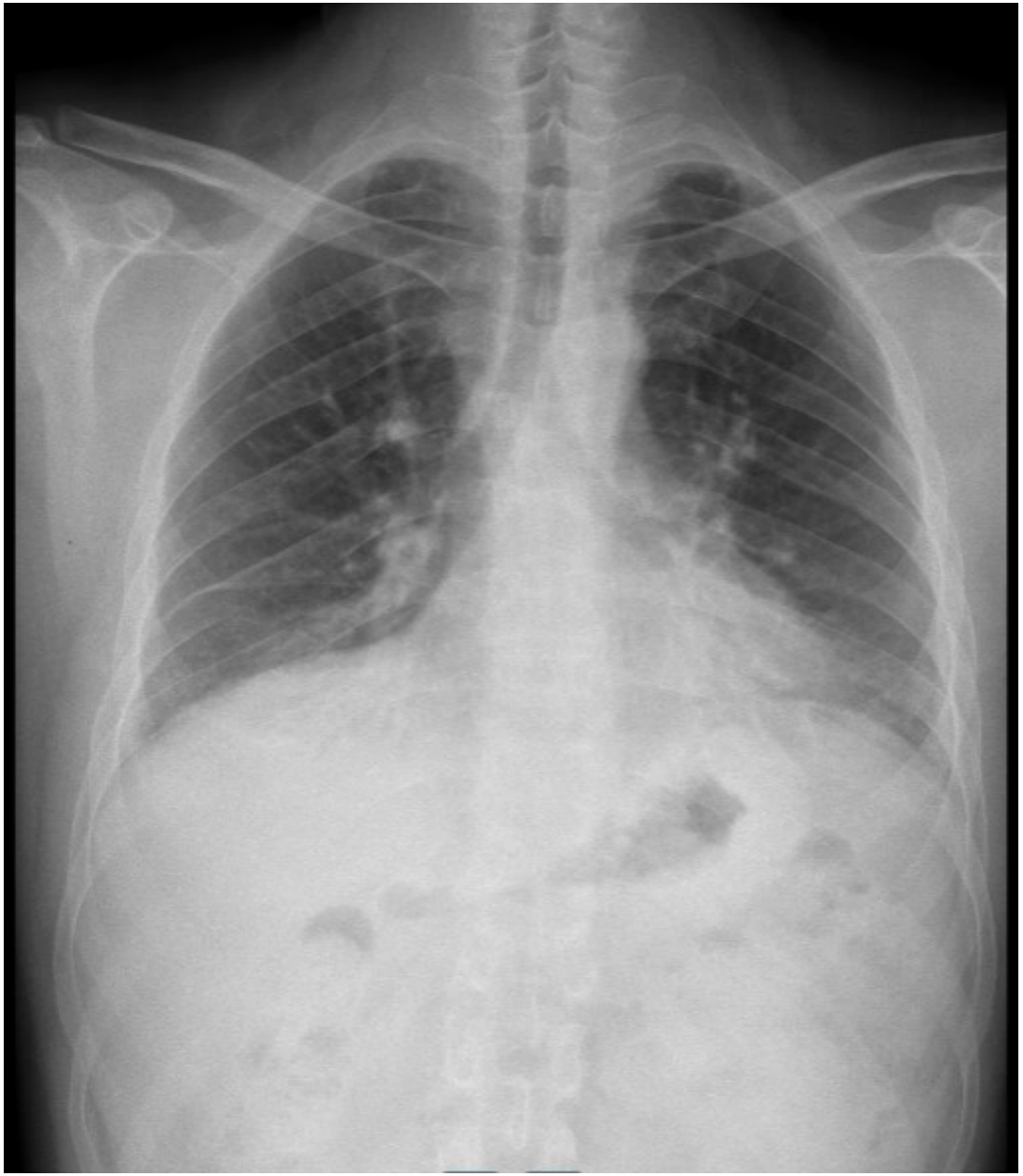
- No pathology was detected in the patient after rhemotology consultation

- Steroids prescribed

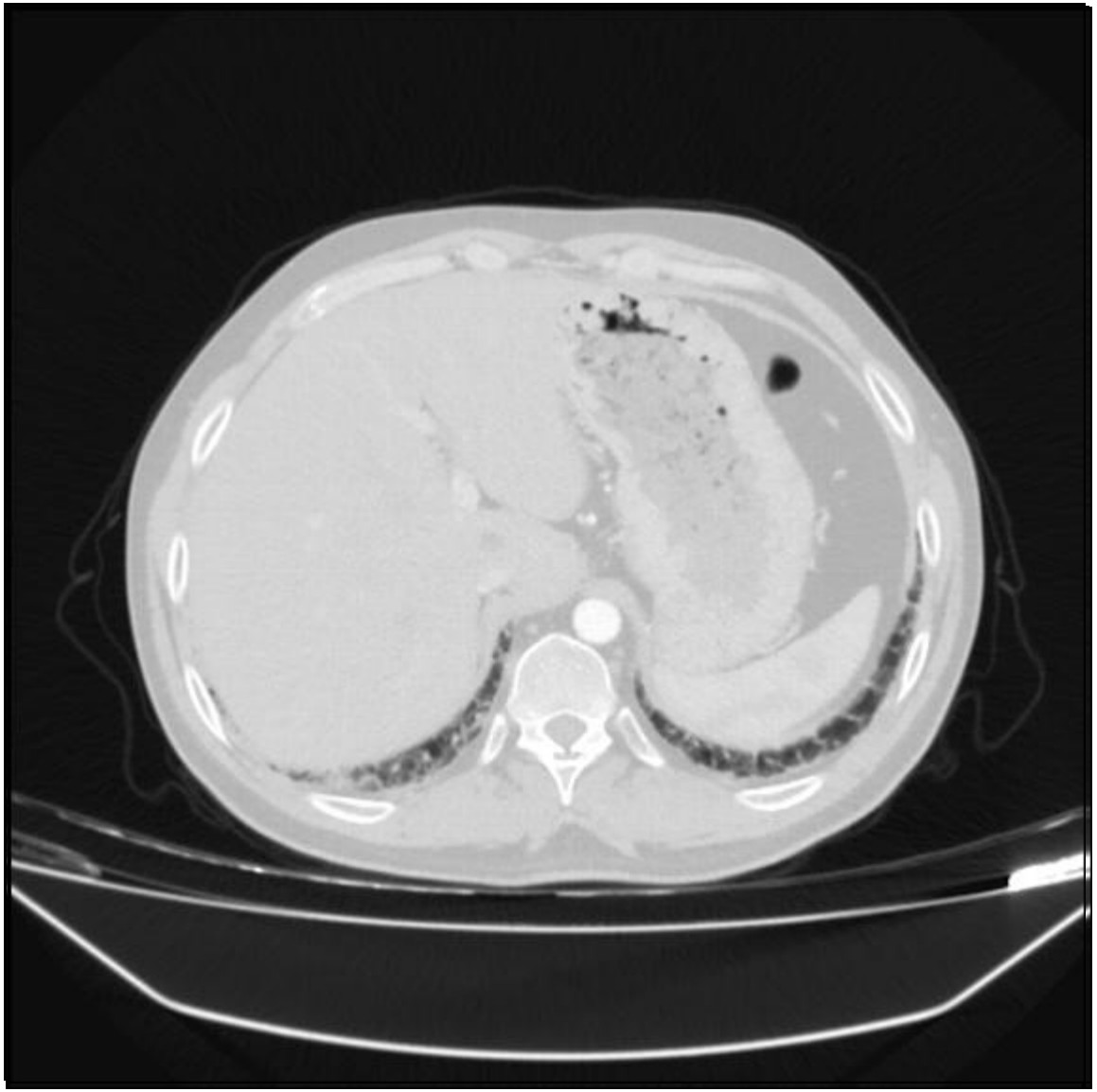
6th month on therapy

	Pre-Bronch Actual	Pred	%Pred	Post-Bronch Actual	%Pred	%Chng
---- SPIROMETRY ----						
FVC (L)	3,10	4,89	63			
FEV1 (L)	2,02	3,93	51			
FEV1/FVC (%)	65	80	82			
FEF 25% (L/sec)	4,90	7,65	64			
FEF 75% (L/sec)	0,27	1,86	15			
FEF 25-75% (L/sec)	0,88	3,83	23			
FEF Max (L/sec)	6,31	9,62	66			
FIVC (L)	2,22					
FIF Max (L/sec)	5,31					

		Ölçüm	Normal Aralık	Bekl.	%Beklenen	z score
DLCO	mL/min/mmHg	**11,07**	25,42 - 39,30	32,36	34	-5,05
DLCO corr	mL/min/mmHg	**11,07**	25,42 - 39,30	32,36	34	-5,05
DLCO/VA	mL/min/mmHg/L	**3,52**	3,41 - 6,17	4,79	73	-1,52
VA	L	**3,15**	5,60 - 7,90	6,75	47	-5,15
TLC(DLCO)	L	**3,30**	5,75 - 8,05	6,90	48	-5,14



13.02.2024



FIRAT ÜNİVERSİTESİ HASTANESİ
RADYOLOJİ RAPORU

Adı Soyadı	:	**Rapor Tarihi**	: 14.02.2024 13:47
T.C Kimlik No	:	**Dosya no**	:
Baba Adı	: NİHAT	**Başvuru No**	:
Kurumu	: SSK SAĞLIK İŞLERİ MÜDÜRLÜĞÜ	**Doğum Yeri - Tarih**	: ELAZIĞ - 1985 **Yaş:** 38
İstem Tarihi	: 13.02.2024(29725199)	**İstem Kabul Tarihi**	: 13.02.2024(R100460)
Hizmet Adi	: BT, TORAKS, KONTRASTLI	**Cinsiyet**	: E

Tanı :	**Kodu**	**Adı**
	R05	ÖKSÜRÜK
	R05	ÖKSÜRÜK

ÇOK KES?TL? B?LG?SAYARLI TOMOGRAF?
TORAKS

Teknik : Kontrast madde verilmeden yap?lan çok kesitli BT tetkikinde;

Trakea ve ana bron?lar, özefagus normal görünümdedir.
Bilateral akci?erlerde orta- alt lobda daha belirgin periferal retikülasyon art??lar? periferal ve paramediastinal subplevral hava kistleri-amfizematöz de?i?iklikler izlendi. Her iki akci?erde yer yer traksiyon bron?ektazileri izlenmektedir (Olas? U?P ?).
Bilateral akci?erde paraseptal amfizematoz de?i?iklikler izlendi.
Orta lobda bant atelektaziler izlendi.
Bilateral akci?er üst lob apikallerde plevroparankimal fibröz kep izlenmektedir.

Peripherical reticulaton on bilateral middle-lower lobes, occasional traction bronchiectasis
(Probable UIP?)

CURRENT MEDICAL RESEARCH AND OPINION
2019, VOL. 35, NO. 11, 2015–2024
https://doi.org/10.1080/03007995.2019.1647040
Article FT-0308.R1/1647040

Taylor & Francis
Taylor & Francis Group

ORIGINAL ARTICLE

Progressive fibrosing interstitial lung diseases: current practice in diagnosis and management

Marlies Wijsenbeek[a], Michael Kreuter[b], Amy Olson[c], Aryeh Fischer[d], Elisabeth Bendstrup[e], Christopher D. Wells[f], Christopher P. Denton[g], Baher Mounir[h], Leila Zouad-Lejour[h], Manuel Quaresma[h] and Vincent Cottin[i]

Table 1. Percentage of US patients who received treatment for non-IPF ILDs in 2016.

	Any treatment[a]	Corticosteroids	Mycophenolate mofetil	Azathioprine	Cyclosporine	Tacrolimus	Cyclophosphamide
RA-ILD	72	69	7	9	5	3	0
SSc-ILD	74	59	29	15	5	4	1
Other CTD-ILDs	67	61	21	15	7	4	0
iNSIP	71	62	15	6	3	3	0
HP	75	74	6	8	2	1	0
Sarcoidosis-ILD	63	62	3	3	2	2	0
Other specified non-IPF ILDs[b]	50	49	3	2	2	1	0
Non-specified ILDs[c]	52	51	3	2	2	2	0

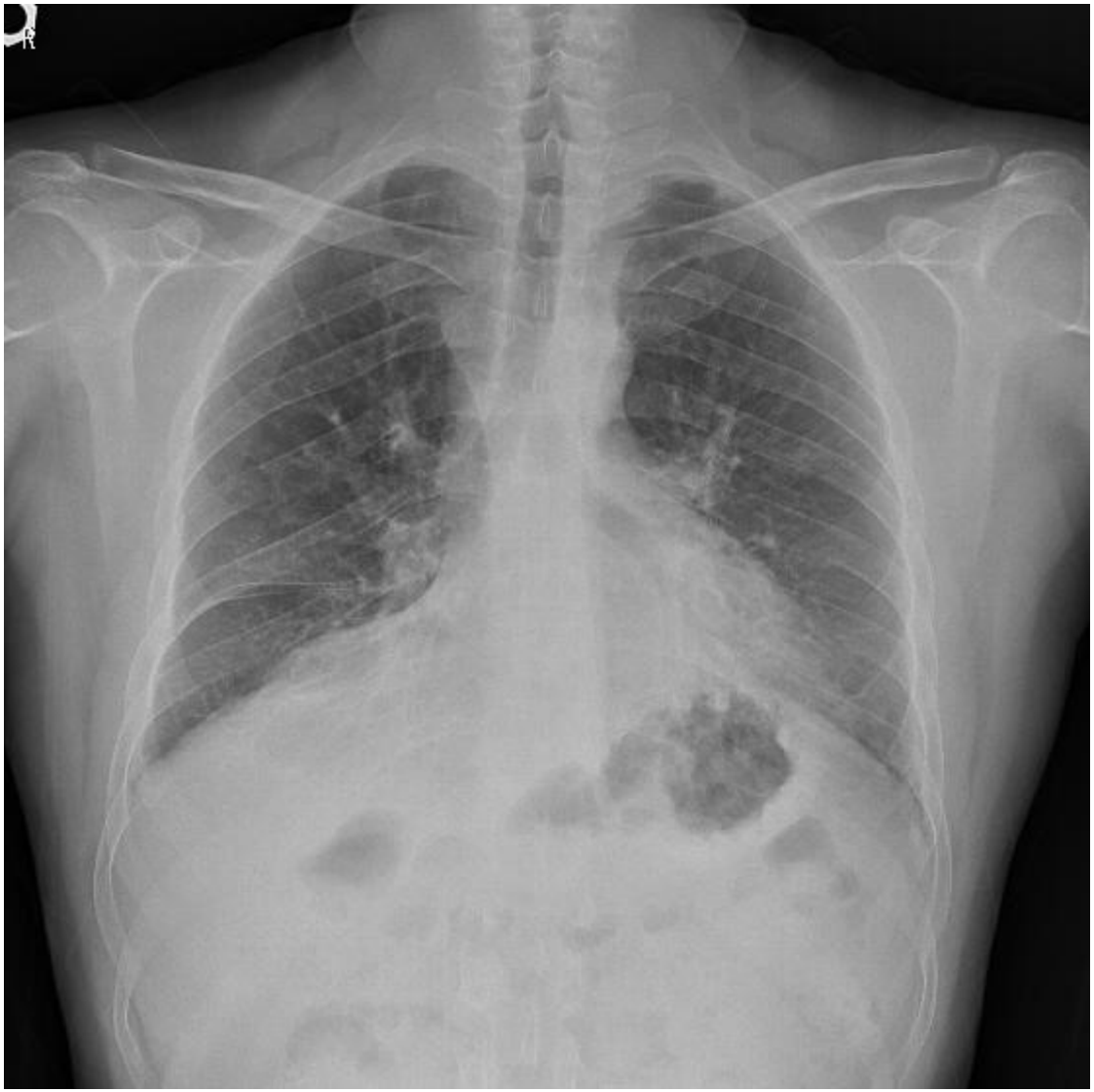
MMF prescribed to the patient

September 2024

- Increase in dyspnea, cough

- PE: Bilaterally inspiratory crackles on middle-lower zones, clubbing (+), **SpO2: %90**

24.09.2024



Name:	[redacted]	ID:	2992	BSA:	1.89	Date:	24.09.2024
Tech:		Height:	173.00	Age:	39	Room:	
Doctor:		Weight:	75.00	Sex:	Male	Race:	Caucasian

Diagnosis:
Dyspnea: Cough: Wheeze:
Tbco Prod: Yrs Smk: Pks/Day: Yrs Quit:
Medications:
Pre Test Comments:
Post Test Comments:

	Pre-Bronch			Post-Bronch		
	Actual	Pred	%Pred	Actual	%Pred	%Chng
---- SPIROMETRY ----						
FVC (L)	1,90	5,00	38			
FEV1 (L)	1,68	4,00	42			
FEV1/FVC (%)	88	80	111			
FEF 25% (L/sec)	4,56	7,61	60			
FEF 75% (L/sec)	1,01	1,83	55			
FEF 25-75% (L/sec)	1,95	3,85	51			
FEF Max (L/sec)	5,31	9,77	54			
FIVC (L)	1,78					
FIF Max (L/sec)	6,21					

		Ölçüm	Normal Aralık	Bekl.	%Beklenen	z score
DLCO	mL/min/mmHg	7,29	25,42 - 39,30	32,36	23	-5,94
DLCO corr	mL/min/mmHg	7,29	25,42 - 39,30	32,36	23	-5,94
DLCO/VA	mL/min/mmHg/L	2,82	3,41 - 6,17	4,79	59	-2,34
VA	L	2,58	5,60 - 7,90	6,75	38	-5,96
TLC(DLCO)	L	2,74	5,75 - 8,05	6,90	40	-5,95

AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Luca Richeldi, Carey C. Thomson, Yoshikazu Inoue, Takeshi Johkoh, Michael Kreuter, David A. Lynch, Toby M. Maher, Fernando J. Martinez, Maria Molina-Molina, Jeffrey L. Myers, Andrew G. Nicholson, Christopher J. Ryerson, Mary E. Strek, Lauren K. Troy, Marlies Wijsenbeek, Manoj J. Mammen, Tanzib Hossain, Brittany D. Bissell, Derrick D. Herman, Stephanie M. Hon, Fayez Kheir, Yet H. Khor, Madalina Macrea, Katerina M. Antoniou, Demosthenes Bouros, Ivette Buendia-Roldan, Fabian Caro, Bruno Crestani, Lawrence Ho, Julie Morisset, Amy L. Olson, Anna Podolanczuk, Venerino Poletti, Moisés Selman, Thomas Ewing, Stephen Jones, Shandra L. Knight, Marya Ghazipura, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This official clinical practice guideline was approved by the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax February 2022

Table 4. Definition of Progressive Pulmonary Fibrosis

Definition of PPF
In a patient with ILD of known or unknown etiology other than IPF who has radiological evidence of pulmonary fibrosis, PPF is defined as at least two of the following three criteria occurring within the past year with no alternative explanation*:
1 Worsening respiratory symptoms
2 Physiological evidence of disease progression (either of the following): a. Absolute decline in FVC ⩾5% predicted within 1 yr of follow-up b. Absolute decline in D_{LCO} (corrected for Hb) ⩾10% predicted within 1 yr of follow-up
3 Radiological evidence of disease progression (one or more of the following): a. Increased extent or severity of traction bronchiectasis and bronchiolectasis b. New ground-glass opacity with traction bronchiectasis c. New fine reticulation d. Increased extent or increased coarseness of reticular abnormality e. New or increased honeycombing f. Increased lobar volume loss

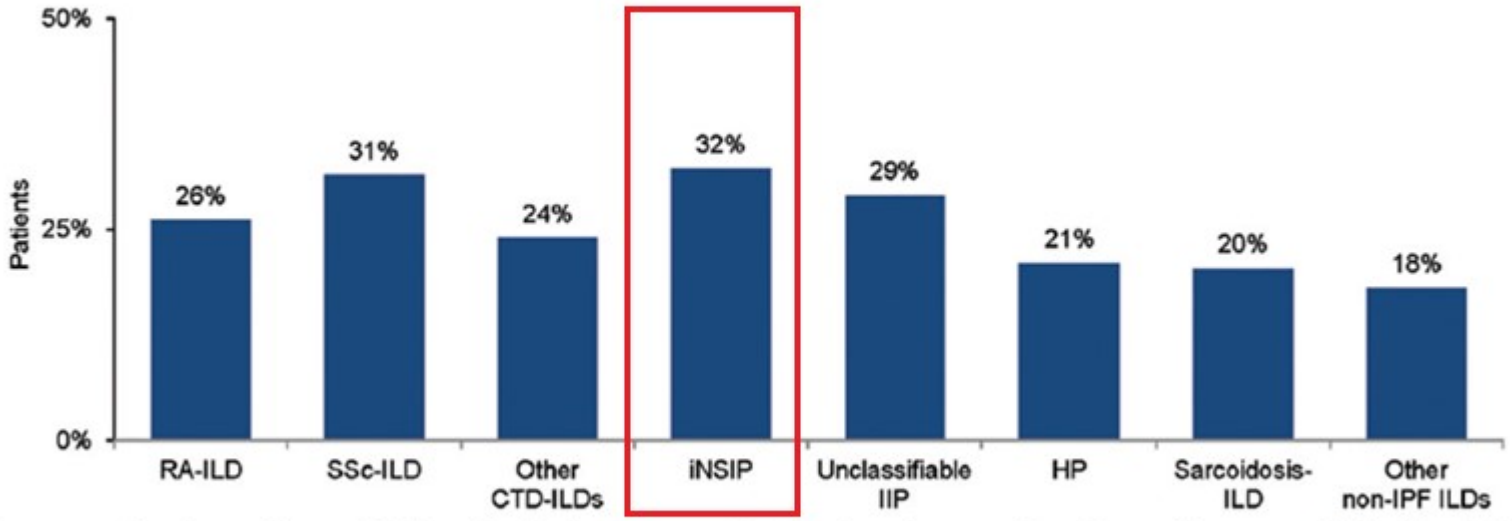


Figure 4. Percentage of patients with non-IPF ILDs who develop a progressive fibrosing phenotype. Data from online survey of physicians (pulmonologists, $n = 243$; rheumatologists, $n = 203$; internists, $n = 40$). Survey question: "For each of the ILD types listed below, among the patients you have seen in the past year, please estimate what percentage of patients had an ILD that (1) had fibrosis detected by HRCT (i.e. reticular abnormality with traction bronchiectasis with or without honeycombing) AND (2) was progressing in terms of worsening of lung function (FVC and/or DL_{CO}) and/or respiratory symptoms and/or chest images". Rheumatologists were only asked this question in relation to RA-ILD, SSc-ILD and other CTD-ILDs. Abbreviations. CTD, Connective tissue disease; DL_{CO}, Diffusing

Wijsenbeek M, et al. Curr Med Res Opin 2019

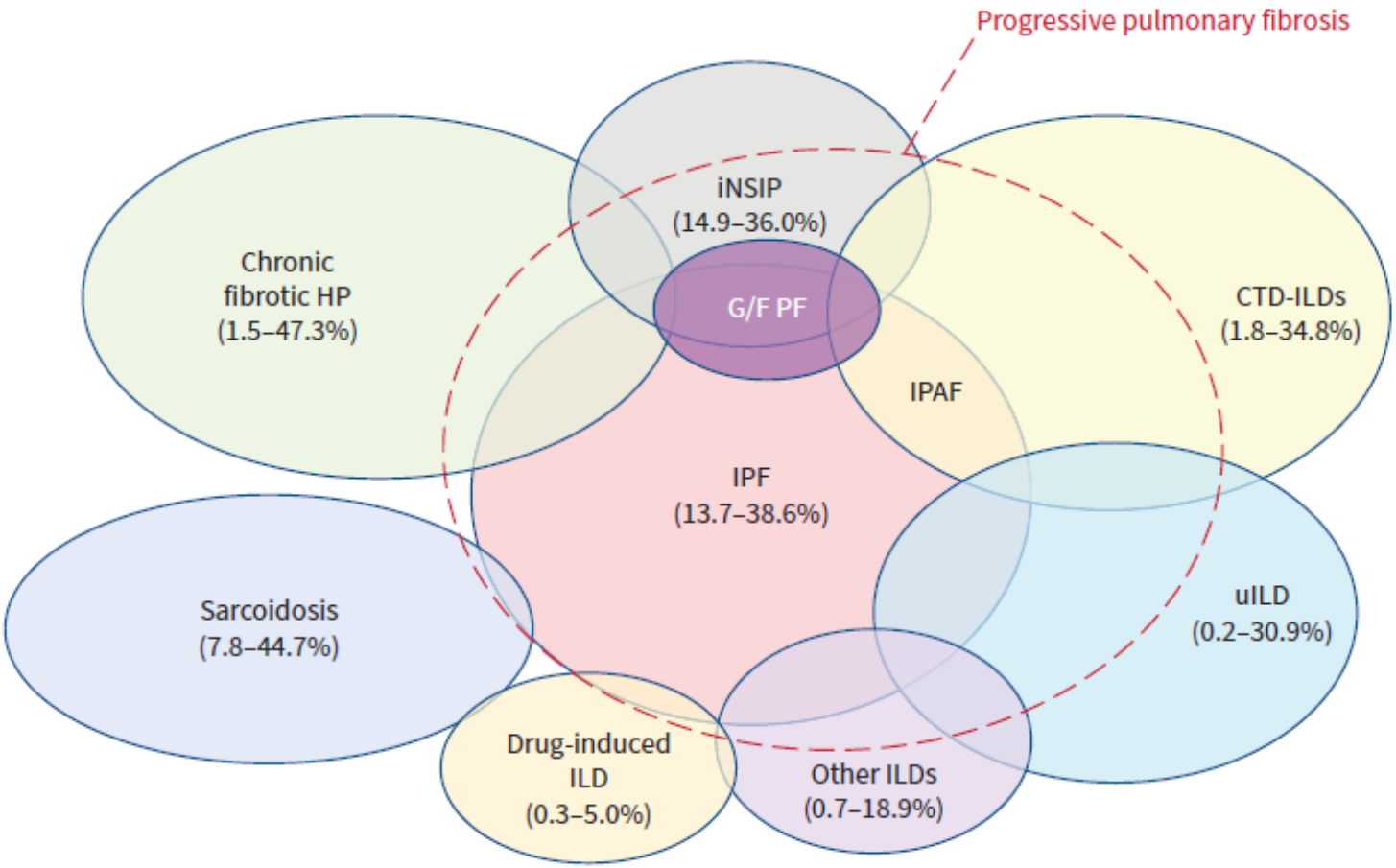


FIGURE 1 Schematic representation of the prevalent spectrum of interstitial lung diseases (ILDs) that may be associated with "progressive pulmonary fibrosis (despite management)". The lowest and highest prevalences across different countries are shown in brackets [14]. CTD: connective tissue disease; G/F PF: genetic and/or

Rajan SK, et al. Eur Respir J Rev 2023

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Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

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Evidence-based Recommendations for Treatment of PPF, Other than IPF

Pirfenidone. ***We recommend further research into the efficacy, effectiveness, and safety of pirfenidone in both 1) non-IPF ILD manifesting PPF in general and 2) specific types of non-IPF ILD manifesting PPF.***

Nintedanib.

We suggest nintedanib for the treatment of PPF in patients who have failed standard management for fibrotic ILD, other than IPF (conditional recommendation, low-quality evidence). *Remarks: Standard*

We recommend research into the efficacy, effectiveness, and safety of nintedanib in specific types of non-IPF ILD manifesting PPF.

Progressive pulmonary fibrosis: an expert group consensus statement

Sujeet K. Rajan [1], Vincent Cottin [2], Raja Dhar[3], Sonye Danoff[4], Kevin R. Flaherty[5], Kevin K. Brown[6], Anant Mohan[7], Elizabeth Renzoni[8], Murali Mohan[9], Zarir Udwadia[10], Padmanabha Shenoy[11], David Currow[12], Anand Devraj[13], Bhavin Jankharia[14], Ritu Kulshrestha[15], Steve Jones[16], Claudia Ravaglia[17], Silvia Quadrelli[18], Rajam Iyer[19], Sahajal Dhooria [20], Martin Kolb [21,23] and Athol U. Wells[22,23]

[1]Bombay Hospital Institute of Medical Sciences and Bhatia Hospital, Mumbai, India. [2]National French Reference Coordinating Center for Rare Pulmonary Diseases, Louis Pradel Hospital Hospices Civils de Lyon, Université Claude Bernard Lyon 1, INRAE, Member of ERN-LUNG, Lyon, France. [3]CK Birla Hospitals, Kolkata, India. [4]Johns Hopkins School of Medicine, Baltimore, MD, USA. [5]University of Michigan, Ann Arbor, MI, USA. [6]Department of Medicine, National Jewish Health, Denver, CO, USA. [7]All India Institute of Medical Sciences, New Delhi, India. [8]Royal Brompton Hospital/Imperial College London, London, UK. [9]Narayana Health, Bengaluru, India. [10]Breach Candy Hospital, Mumbai, India. [11]Department of Rheumatology, Centre for Arthritis and Rheumatism Excellence, Kochi, India. [12]University of Technology, Sydney, Australia. [13]Department of Radiology, Royal Brompton Hospital, London, UK. [14]Picture This by Jankharia, Mumbai, India. [15]Department of Pathology, Vallabhbhai Patel Chest Institute, University of Delhi, Delhi, India. [16]European Idiopathic Pulmonary Fibrosis Federation (EU-IPFF), Peterborough, UK. [17]Pulmonology Unit, GB Morgagni Hospital/ University of Bologna, Forlì, Italy. [18]Hospital Britanico de Buenos Aires, Buenos Aires, Argentina. [19]Bhatia Hospital and PD Hinduja Hospital, Mumbai, India. [20]Department of Pulmonary Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, India. [21]Firestone Institute for Respiratory Heath, St Joseph's Healthcare and McMaster University, Hamilton, ON, Canada. [22]Interstitial Lung Disease Unit, Royal Brompton and Harefield NHS Foundation Trust, London, UK. [23]Co-senior authors.

Corresponding author: Sujeet K. Rajan (skrajan@hotmail.com)

Shareable abstract (@ERSpublications)
Progressive pulmonary fibrosis (PPF) explains what clinicians increasingly face in practice. Assessing ILD progression, its risk and improved treatments based on current evidence for PPF (despite initial management) form the mainstay of this document. http://bit.ly/3GLdqfs

Cite this article as: Rajan SK, Cottin V, Dhar R, *et al*. Progressive pulmonary fibrosis: an expert group consensus statement. *Eur Respir J* 2023; 61: 2103187 [DOI: 10.1183/13993003.03187-2021].

Key conclusions

1) The long-term side effects with systemic corticosteroid therapy are a cause for concern in the treatment of PPF; alternative long-term immunosuppressive agents may be associated with less side effects [77].
2) A case-by-case and disease-by-disease approach and review are required to assess the added effectiveness of immunosuppressants to the baseline steroid treatment.

Key conclusions

1) Initial treatment should be based on the precise primary diagnosis.
2) Apart from IPF, SSc-ILD and, possibly, rheumatoid arthritis-associated ILD (UIP), antifibrotic medication should not be considered as a first-line therapy.
3) In PPF, there is growing evidence that antifibrotic therapy reduces lung function decline, regardless of background immunosuppressive therapy.
4) Careful monitoring for adverse events in the patient subgroup treated with combination therapy is advised.

Key conclusions

1) The group does not advise general upfront combination therapy.
2) Antifibrotics may be administered sequentially in the context of PPF.

Tanı ve İlaç Bilgileri

Tanı Kodu	Tanı Adı	Alt Tanı	Tanı Tarihi
J84.1	İntersitisyel akciğer hastalığı, diğer, fibrozisli	fibrotik NSİP	24.09.2024

Etkin Madde	Miktarı	Doz 1	X	Doz 2	Periyodu	K. Şekli	Rapor Süresi
PIRFENIDON	600 mg	4	X	1	1 Gün	Ağızdan(Oral)	1 Yıl

Açıklama

İlgili etkin maddeli ilacın kullanımı, Kısa Ürün Bilgisi (KÜB) dahilinde olup, 1 (bir) yıllık dozda kullanımı uygundur.

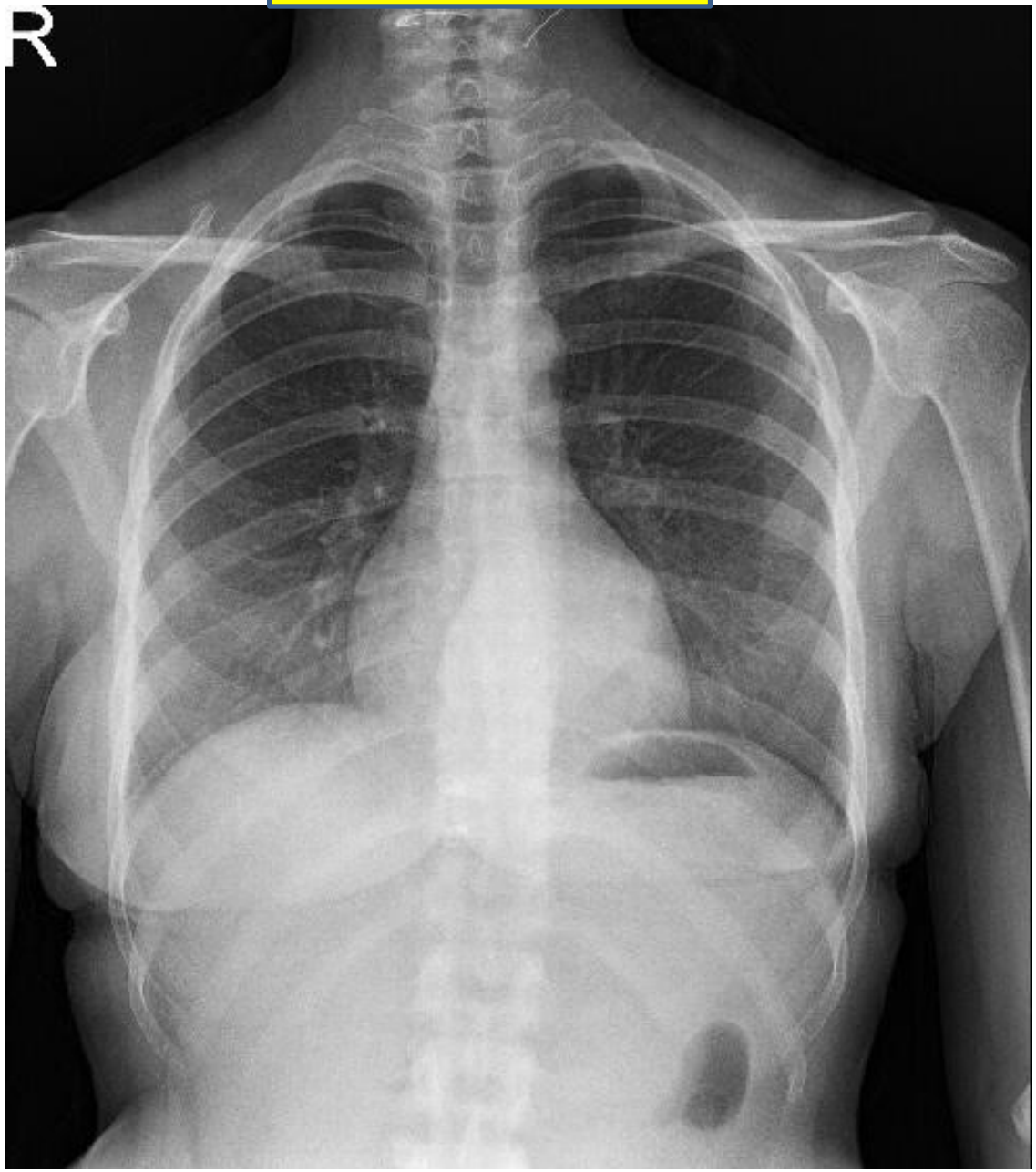
Karar	Karar Tarihi
Uygun görülmüştür	26.09.2024

He was started on **pirfenidone** with off-label approval

Case-2

- 34 y, F
- She applied to the Rhematology outpatient clinic 5 years ago due to joint pain
- PE: No pathological sign
- No laboratory exam

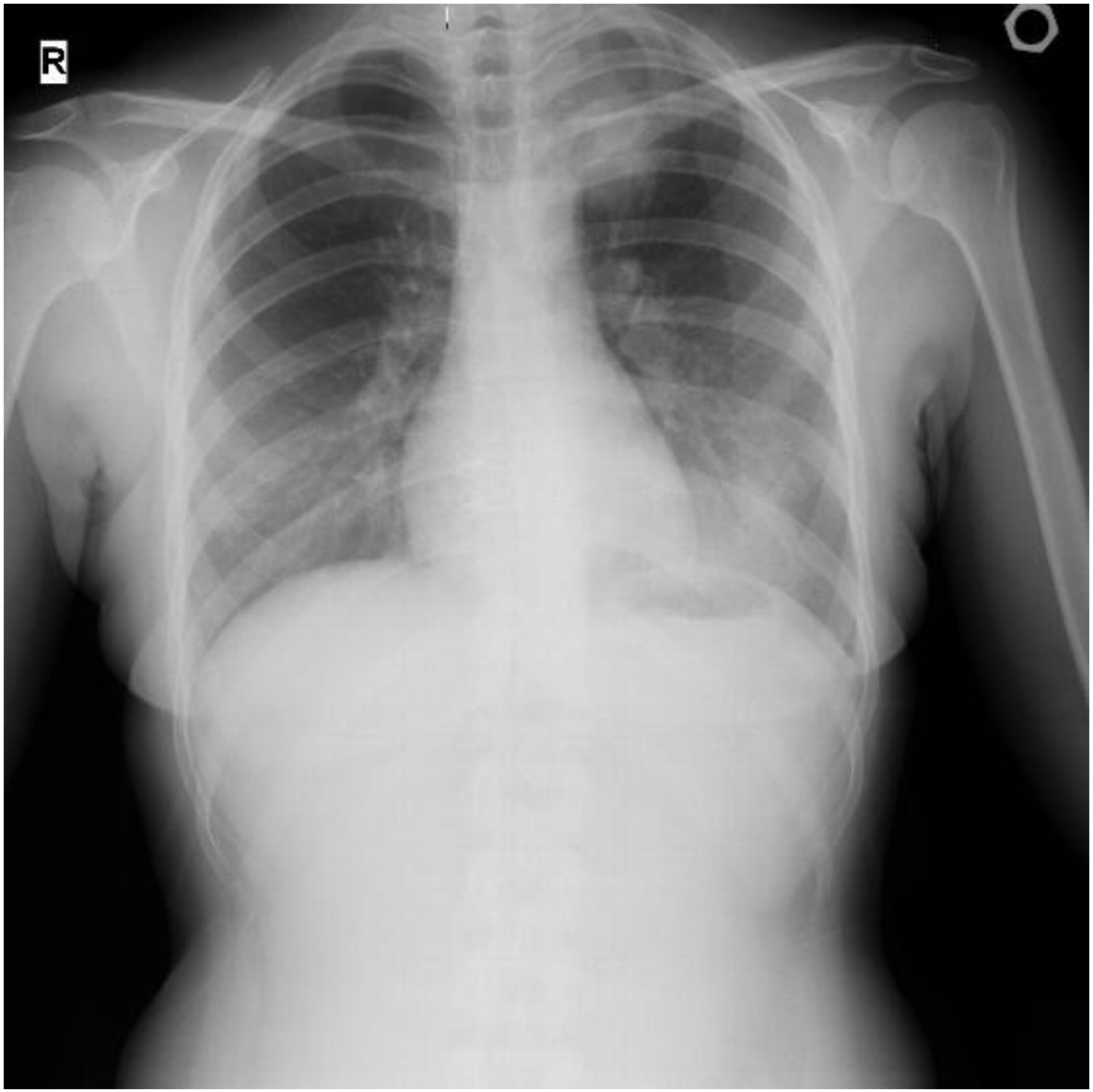
01.07.2019



- Anti-inflammatory treatment

- Re-applied to the Rhematology outpatient clinic after 3 months because of persistant joint pain

23.10.2019



Laboratory findings

- **Hemogram**
  - ✓ WBC: 8340/µL
  - ✓ **Hb:10.7** g/dL
  - ✓ **Hct: 32 %**
  - ✓ Plt: 435000/µL
  - ✓ **CRP: 200 mg/L** (0-5)

- **Biochemistry**
  - ✓ Urea: 29 mg/dl
  - ✓ Cr:0.58 mg/dl
  - ✓ AST: 37 U/L
  - ✓ **ALT: 76 U/L**
  - ✓ **Urine analysis**
    - **+++** Ery/uL

☑	23.10.2019 08:50	MONOKLONAL ANTİKOR (HER BİRİ)	Negatif		null - null
☑	23.10.2019 08:50	CYCLİC CİTRULLİNATED PEPTİDE (CCP)	1,1	u/mL	0 - 5
☑	23.10.2019 08:50	Romatoid faktör (RF) (Nefelometrik)	104	IU/mL	0 - 15
☑	23.10.2019 08:50	VİTAMİN B12	404	pg/mL	174 - 878
☑	23.10.2019 08:50	FERRİTİN	1004.5	ng/mL	null - null
☑	23.10.2019 08:50	DEMİR BAĞLAMA KAPASİTESİ	281	ug/dL	250 - 450
☑	23.10.2019 08:50	Demir (Serum)	7	ug/dL	60 - 170

Reçete İçin Seçilmiş ICD 10 Tanıları

Tanı Kodu	Tanı Adı
M36.8	BAĞ DOKUSUNUN SİSTEMİK BOZUKLUKLARI, BAŞKA YERDE SINIFLANMIŞ DİĞER

Systemic Disorders of Connective Tissue

K21.9	GASTRO-ÖZOFAJİAL REFLÜ HASTALIĞI, ÖZOFAJİTSİZ
M79.7	FİBROMİYALJİ

Hasta Adı :
TC Kimlik No :
Reçete S.No : 2075131
Takip No : 31GBB9L

Reçete Tarih : 23.10.2019 00:00:00
Reçete Türü : Normal
Reçete Tipi : Ayaktan Reçetesi
Reçete No : 18B3VTL

Reçeteyi Yazan Doktor :
Branş Kodu : ROMATOLOJİ

Barkod	Ilaç Adı	Adet	Doz	Periyodu	Periyot Birim	Kullanım Sekli
8699638012053	EMTHEXATE 2.5 MG 100 TB.	,0	1,00 X 6,00	1,00	Hafta	Ağızdan(Oral)
8699532010865	DELTACORTRIL 20 TABLET	5,0	1,00 X 3,00	1,00	Gün	Ağızdan(Oral)

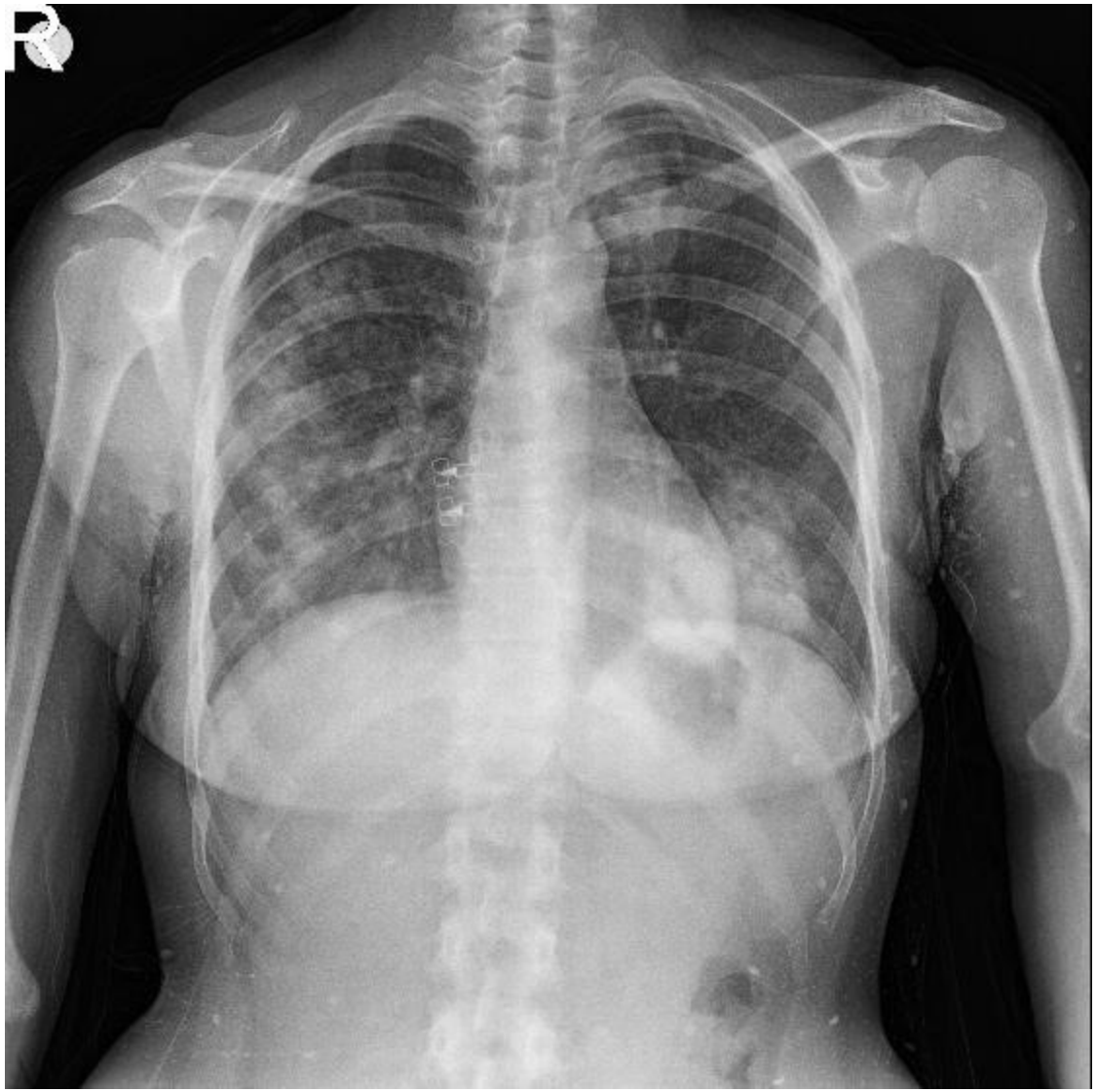
After 2 weeks

- Applying to the emergency department with complaints of shortness of breath and **hemoptysis**
- PE: Conscious, cooperated, oriented
- Chest oscultation: Bilaterally inspiratory crackles
- **SpO2: %90**, TA: 110/70 mmHg, **Pulse: 134/dk**, **RR: 26/dk**, Fever: 37.2C

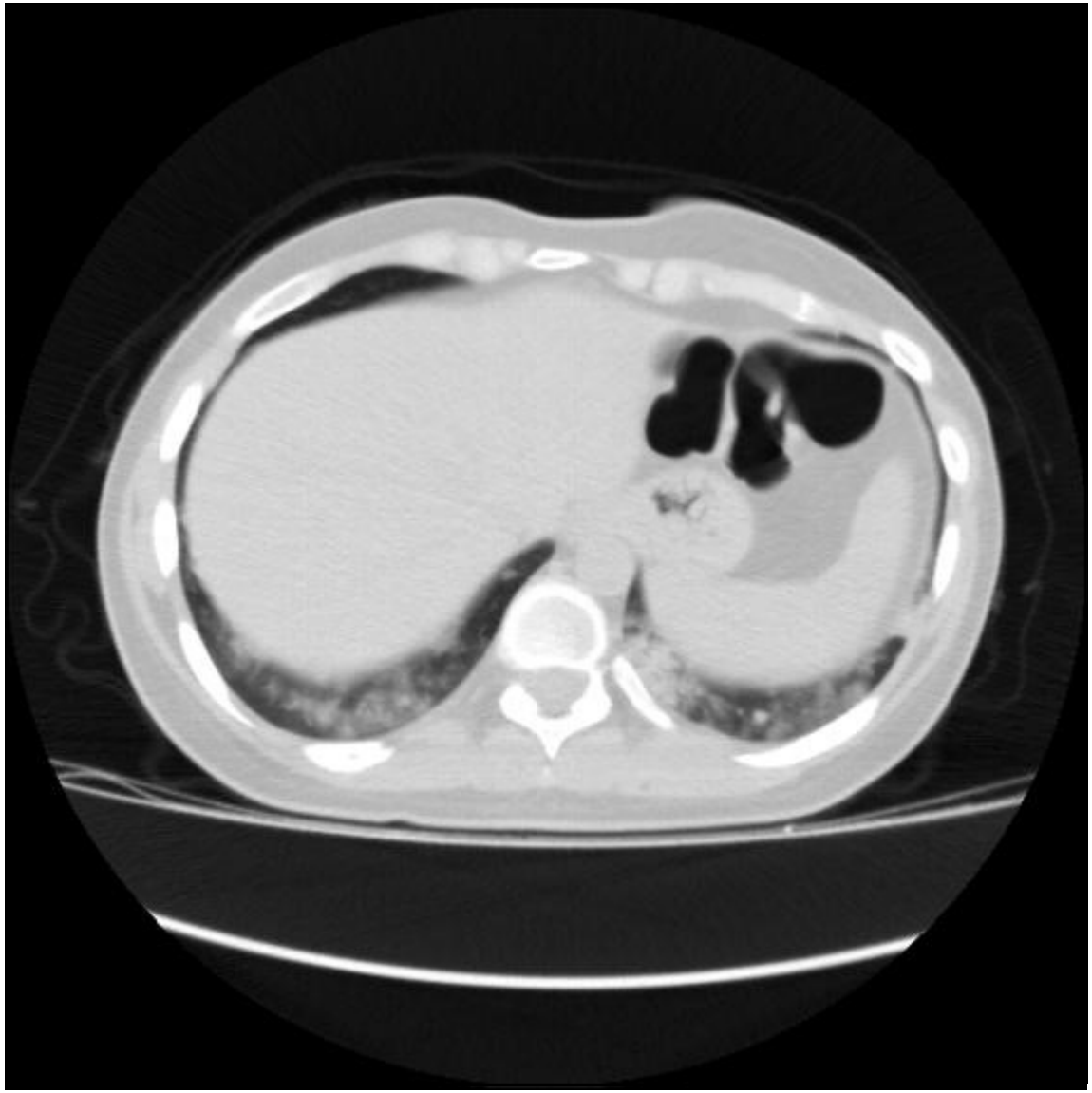
Laboratory findings

- Hemogram
  - ✓ **WBC: 9410/µL**
  - ✓ **Hb:8.5** g/dL
  - ✓ **Hct: 26.6 %**
  - ✓ Plt: 374000/µL
  - ✓ **CRP: 145 mg/L** (0-5)
- Biochemistry
  - ✓ **Urea: 52 mg/dl**
  - ✓ Cr:0.58 mg/dl
  - ✓ AST: 37 U/L
  - ✓ **ALT: 49 U/L**
  - ✓ Urine analysis
    - **+++** Ery/uL

09.11.2019



12.11.2019



T.C.
FIRAT ÜNİVERSİTESİ HASTANESİ
RADYOLOJİ RAPORU

Adı Soyadı	:	**Rapor Tarihi**	: 12.11.2019 01:20
T.C Kimlik No	:	**Dosya no**	:
Baba Adı	: ŞÜKRÜ	**Başvuru No**	:
Kurumu	: SSK SAĞLIK İŞLERİ MÜDÜRLÜĞÜ	**Doğum Yeri - Tarih**	: ELAZIĞ - 1990 **Yaş:** 29
İstem Tarihi	: 12.11.2019(18295976)	**İstem Kabul Tarihi**	: 12.11.2019(46261)
Hizmet Adi	: BT, TORAKS	**Cinsiyet**	: K

Tanı :	**Kodu**	**Adı**
	R07.4	GÖĞÜS AĞRISI, TANIMLANMAMIŞ
	R06.0	DİSPNE
	M79.7	FİBROMİYALJİ
	D64	ANEMİ, DİĞER
	R06	SOLUNUM ANORMALLİKLERİ
	M25.5	EKLEM AĞRISI
	M79.9	YUMUŞAK DOKU BOZUKLUĞU, TANIMLANMAMIŞ
	R77.0	ALBÜMİN ANORMALLİĞİ
	M31.3	WEGENER GRANÜLOMATOZU

AC?L ÇOK KES?TL? B?LG?SAYARLI TOMOGRAF?
TORAKS

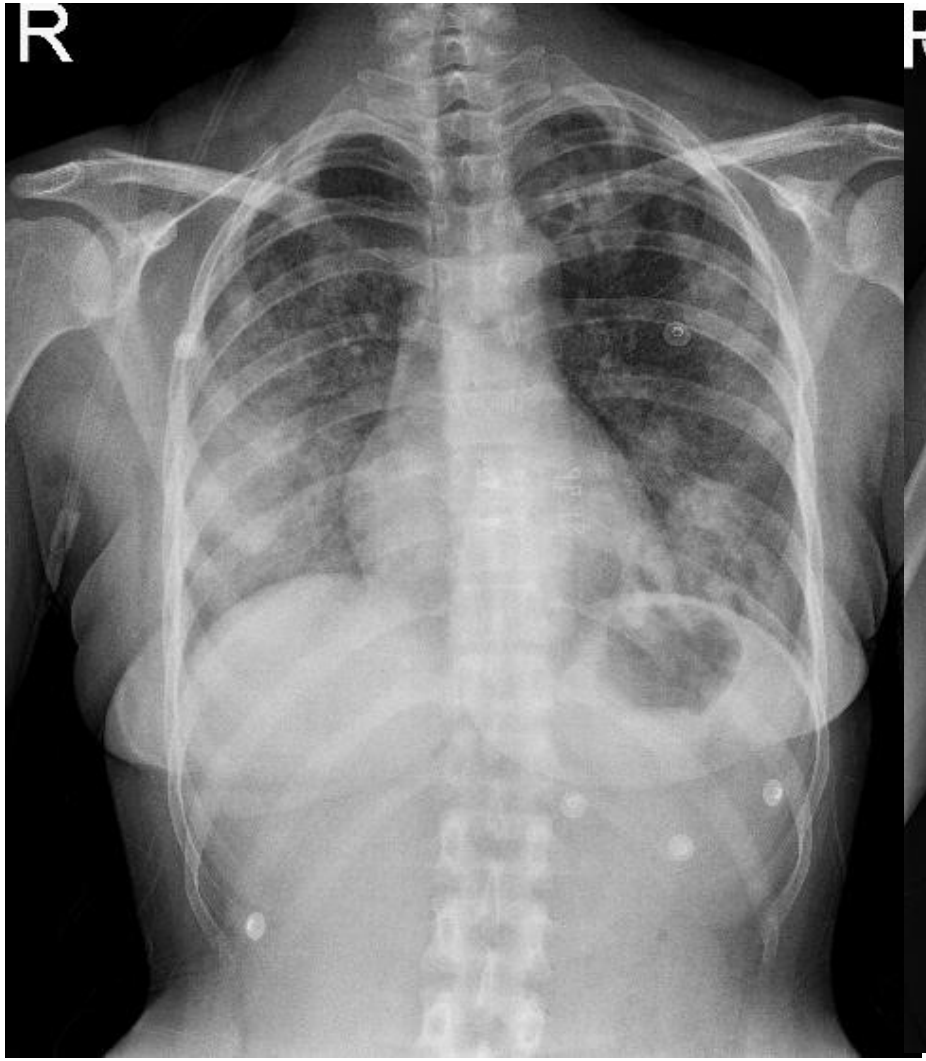
Teknik : Kontrast madde verilmeden yap?lan 64-ÇKBT incelemesinde:

Bilateral akci?erde yayg?n yamal? konsolidasyon alanlar?, tomurcuklanm?? a?aç görünümleri ve yer yer kavitelerinde izlendi?i görünüm dikkati çekmektedir (Akci?er tüberkülozu ?Vaskülit ?).

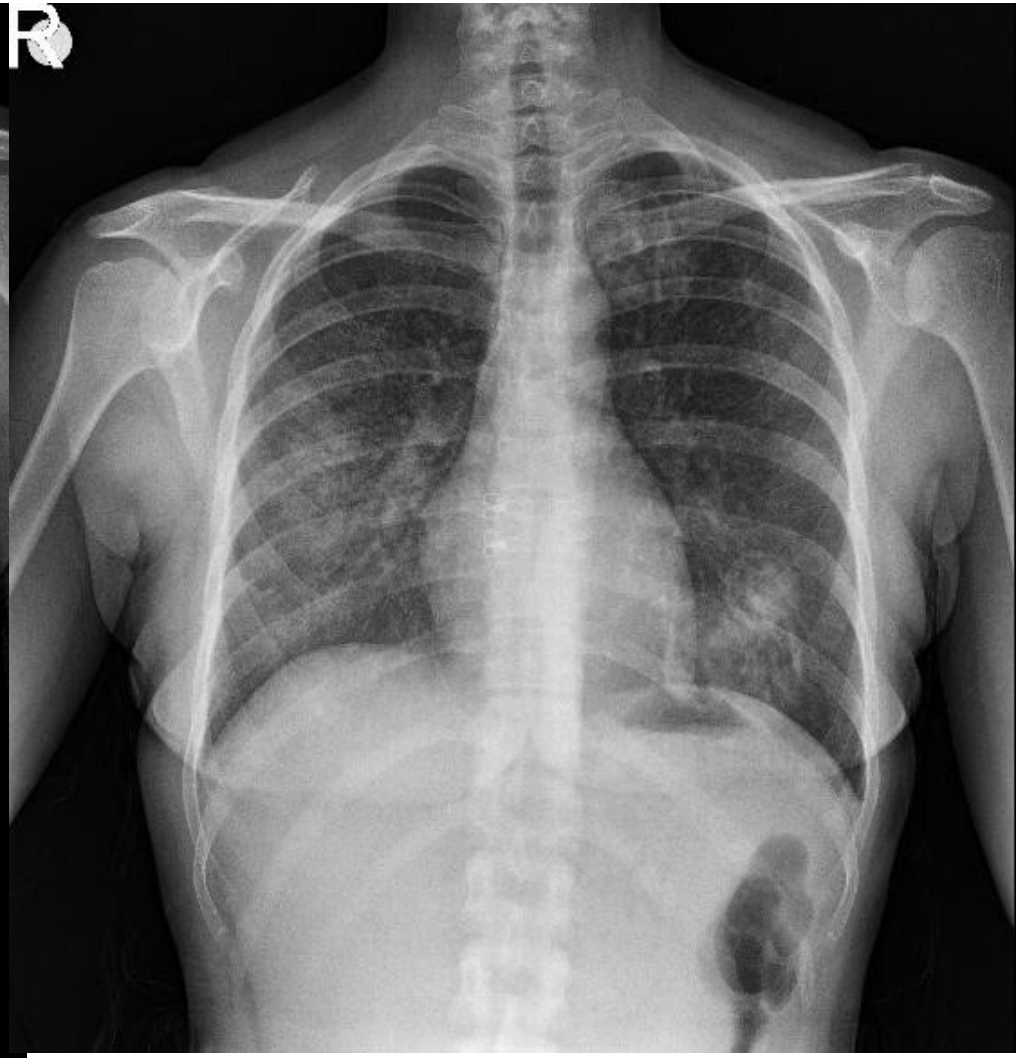
Thorax CT: Bilaterally patchy consolidation, tree-in bud pattern, partly cavitation (Tuberculosis? Vasculitis?)

- She hospitalized to the ICU with alveolar hemorrhage diagnosis
- 1 gr Metilprednizolon for 3 days, after then 1 mg/kg (IV)
- Cyclophosphamide 500 mg IV
- Apheresis
- Rituximab prescribed after 2 weeks

13.11.2019



18.11.2019



FIRAT ÜNİVERSİTESİ HASTANESİ
RADYOLOJİ RAPORU

Adı Soyadı	:	**Rapor Tarihi**	: 14.11.2019 14:48
T.C Kimlik No	:	**Dosya no**	:
Baba Adı	: ŞÜKRÜ	**Başvuru No**	:
Kurumu	: SSK SAĞLIK İŞLERİ MÜDÜRLÜĞÜ	**Doğum Yeri - Tarih**	: ELAZIĞ - 1990 **Yaş:** 29
İstem Tarihi	: 12.11.2019(18301962)	**İstem Kabul Tarihi**	: 12.11.2019(46255)
Hizmet Adi	: BT, PARANAZAL SINÜS	**Cinsiyet**	: K

Tanı :	**Kodu**	**Adı**
	R07.4	GÖĞÜS AĞRISI, TANIMLANMAMIŞ
	R06.0	DİSPNE
	M79.7	FİBROMİYALJİ
	D64	ANEMİ, DİĞER
	R06	SOLUNUM ANORMALLİKLERİ
	M25.5	EKLEM AĞRISI
	M79.9	YUMUŞAK DOKU BOZUKLUĞU, TANIMLANMAMIŞ
	R77.0	ALBÜMİN ANORMALLİĞİ
	M31.3	WEGENER GRANÜLOMATOZU

KL?N?K: Wegener vaskülit

PARANAZAL S?NÜS BT ?NCELEMES?

Koronal planda al?nan kesitlerin de?erlendirilmesinde

Bilateral orta konkada konka bülloza ile uyumlu görünüm izlendi.
Sa? maksiller sinüste ve sfenoid sinüs sol komponentte mukozal kal?nla?malar izlenmektedir (?nflamatuar Sinüs Hastal???).
Sol maksiller sinusun aerasyonlar? tabiidir.

Paranasal CT: Inflammatory sinus disease

PATOLOJİ RAPORU

BİYOPSİ RAPORU

Hasta Adı Soyadı	-	Biyopsi/Sitoloji No	B-16164/19
Biyopsi/Sitoloji No	**B-16164/19**	Isteyen Bölüm	ROMATOLOJİ YOĞUN BAKIM ÜNİTESİ
T.C. Kimlik No	4562*******	Tetkiki Isteyen Doktor	
Yaş / Cinsiyeti	29 / K	Rapor Ilk Kayıt Tarihi	
DosyaNo	809719		
Tetkik Istem Zamanı	21.11.2019 11:04	Numune Kabul Zamanı	21.11.2019 11:08
Numune Alma Zamanı	21.11.2019 11:04	Uzman Onay Zamanı	02.01.2020 09:54
Rapor Onay Tarihi	30.12.2019 10:25	Istem Tarihi	21.11.2019(18380092
Rapor Kes. Tarihi	02.01.2020 09:54	Istem Kabul Tarihi	21.11.2019(909520)

KLİNİK ÖZET:

KLİNİK BİLGİ:

KL?N?K ÖN TANI: Wegener granülomatozu?

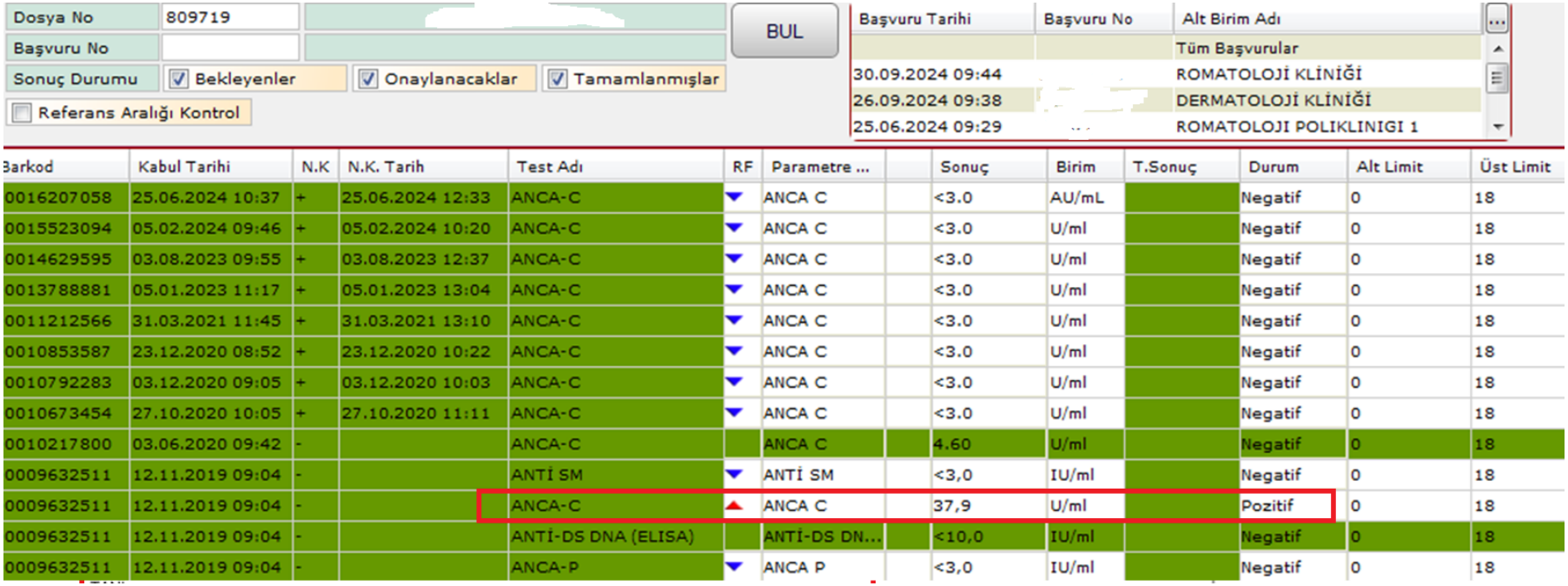
MAKROSKOB?: Formol tespitli 0,3-0,2 cm çap?nda kirli beyaz renkli düzensiz ?ekilli 2 adet materyalin tümü takibe al?nd? (2P-1K)

H?STOK?MYASAL BOYALAR:
PAS:Ola?an
GMS:Ola?an

TANI:
BURUN SA? SEPTUM MUKOZASI, PUNCH B?YOPS?:
-ÜLSER, ?LT?HAB? GRANÜLASYON DOKUSU VE AKT?F KRON?K ?NFLAME MUKOZAL DOKU FRAGMANLARI

Yorum: Gönderilen biyopsi örnekleri nonspesifik ülsere yüzeyel mukozal doku fragmanlar?ndan ibaret olup mevcut biyopsiyle klinik ön tan?ya yönelik daha ileri yorum yap?lamam??t?r. Klinik korelasyon önerilir.

Nasal septum mucosa biopsy: Ulcer, inflammatory granulation, active chronic mucosal tissue



Dosya No 809719

Başvuru No

Sonuç Durumu ☑ Bekleyenler ☑ Onaylanacaklar ☑ Tamamlanmışlar

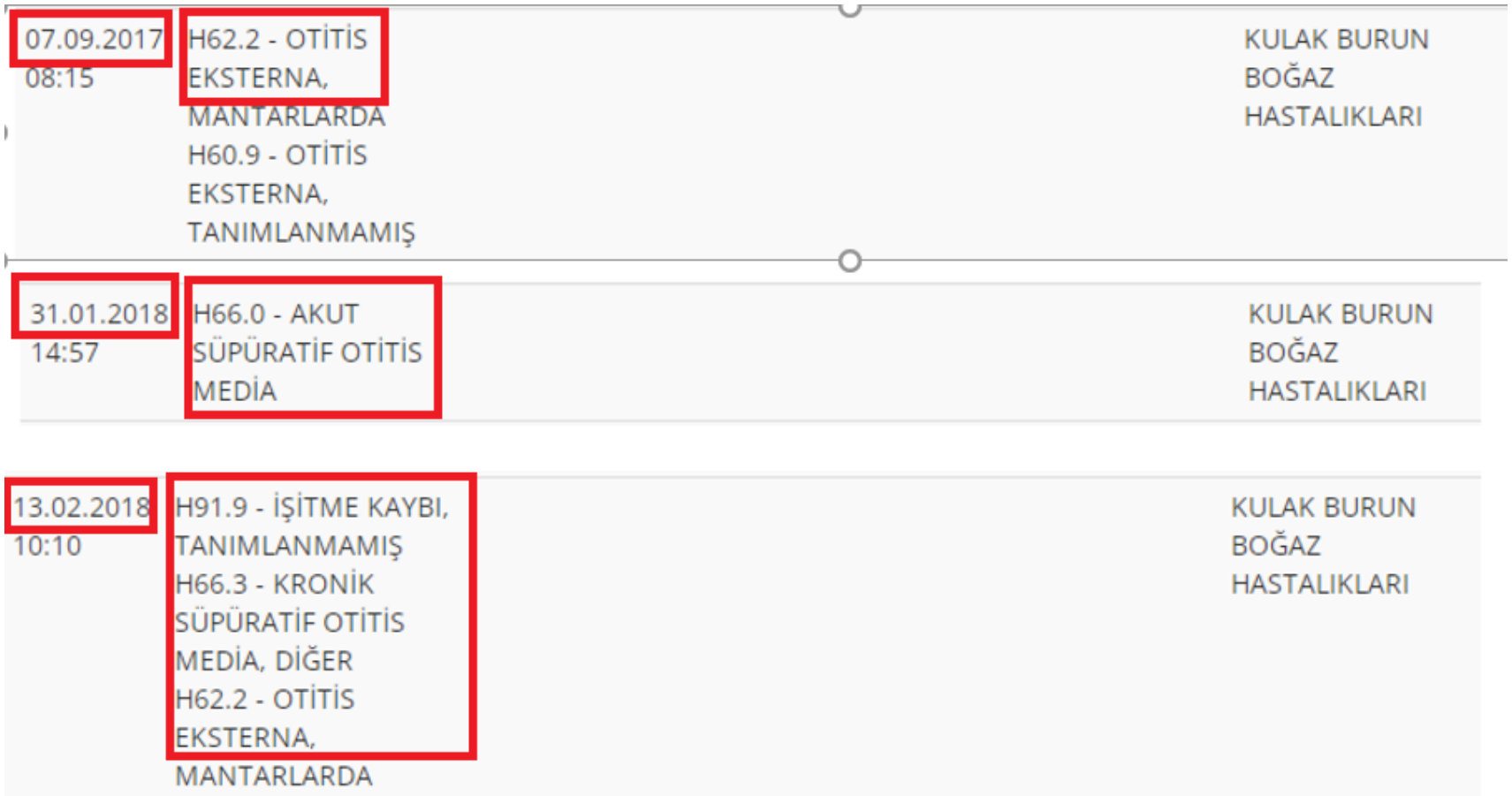
☐ Referans Aralığı Kontrol

BUL

Başvuru Tarihi	Başvuru No	Alt Birim Adı
		Tüm Başvurular
30.09.2024 09:44		ROMATOLOJİ KLİNİĞİ
26.09.2024 09:38		DERMATOLOJİ KLİNİĞİ
25.06.2024 09:29		ROMATOLOJI POLIKLINIGI 1

Barkod	Kabul Tarihi	N.K	N.K. Tarih	Test Adı	RF	Parametre ...		Sonuç	Birim	T.Sonuç	Durum	Alt Limit	Üst Limit
0016207058	25.06.2024 10:37	+	25.06.2024 12:33	ANCA-C	▼	ANCA C		<3.0	AU/mL		Negatif	0	18
0015523094	05.02.2024 09:46	+	05.02.2024 10:20	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0014629595	03.08.2023 09:55	+	03.08.2023 12:37	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0013788881	05.01.2023 11:17	+	05.01.2023 13:04	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0011212566	31.03.2021 11:45	+	31.03.2021 13:10	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0010853587	23.12.2020 08:52	+	23.12.2020 10:22	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0010792283	03.12.2020 09:05	+	03.12.2020 10:03	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0010673454	27.10.2020 10:05	+	27.10.2020 11:11	ANCA-C	▼	ANCA C		<3.0	U/ml		Negatif	0	18
0010217800	03.06.2020 09:42	-		ANCA-C		ANCA C		4.60	U/ml		Negatif	0	18
0009632511	12.11.2019 09:04	-		ANTİ SM	▼	ANTİ SM		<3,0	IU/ml		Negatif	0	18
0009632511	12.11.2019 09:04	-		ANCA-C	▲	ANCA C		37,9	U/ml		Pozitif	0	18
0009632511	12.11.2019 09:04	-		ANTİ-DS DNA (ELISA)		ANTİ-DS DN...		<10,0	IU/ml		Negatif	0	18
0009632511	12.11.2019 09:04	-		ANCA-P	▼	ANCA P		<3,0	IU/ml		Negatif	0	18

Past medical history: Otitis externa, otitis media, hearing loss



07.09.2017 08:15	H62.2 - OTİTİS EKSTERNA, MANTARLARDA H60.9 - OTİTİS EKSTERNA, TANIMLANMAMIŞ	KULAK BURUN BOĞAZ HASTALIKLARI
31.01.2018 14:57	H66.0 - AKUT SÜPÜRATİF OTİTİS MEDİA	KULAK BURUN BOĞAZ HASTALIKLARI
13.02.2018 10:10	H91.9 - İŞİTME KAYBI, TANIMLANMAMIŞ H66.3 - KRONİK SÜPÜRATİF OTİTİS MEDİA, DİĞER H62.2 - OTİTİS EKSTERNA, MANTARLARDA	KULAK BURUN BOĞAZ HASTALIKLARI

Ann Rheum Dis. 2022 Mar;81(3):315-320. doi: 10.1136/annrheumdis-2021-221795.
Epub 2022 Feb 2.

2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis

Joanna C Robson [1], Peter C Grayson [2], Cristina Ponte [3], Ravi Suppiah [4], Anthea Craven [5], Andrew Judge [6 7], Sara Khalid [5], Andrew Hutchings [8], Richard A Watts [5 9], Peter A Merkel [10], Raashid A Luqmani [5]; DCVAS Investigators

- Bloody nasal discharge, nasal crusting, or sinonasal congestion (+3)
- Cartilaginous involvement (+2)
- Conductive or sensorineural hearing loss (+1)
- Cytoplasmic ANCA or anti-PR3 ANCA positivity (+5)
- Pulmonary nodules, mass, or cavitation on chest imaging (+2)
- Granuloma or giant cells on biopsy (+2)
- Inflammation or consolidation of the nasal/paranasal sinuses on imaging (+1)
- Pauci-immune glomerulonephritis (+1)
- Perinuclear ANCA or anti-MPO ANCA positivity (−1)
- Eosinophil count more than 1×10^9 cells/L (−4)

Total score: 10

After excluding mimics of vasculitis, a patient diagnosed with small- or medium-vessel vasculitis could be classified as having GPA if the cumulative score is 5 or more points. When these criteria were tested in the validation dataset, the sensitivity was 93%, and the specificity was 94%.[28]

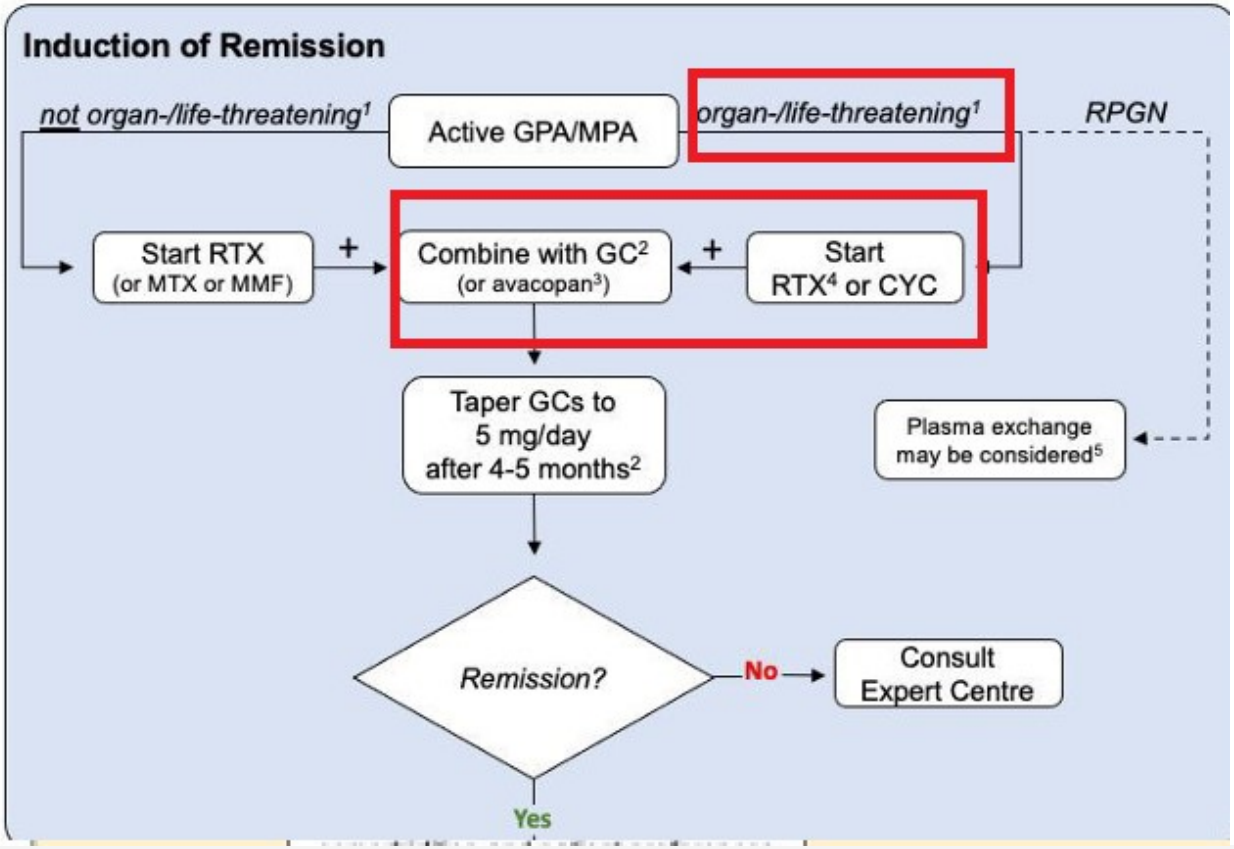
EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update

Bernhard Hellmich ,[1] Beatriz Sanchez-Alamo,[2] Jan H Schirmer,[3] Alvise Berti ,[4,5] Daniel Blockmans,[6] Maria C Cid ,[7] Julia U Holle,[8] Nicole Hollinger,[1] Omer Karadag,[9] Andreas Kronbichler,[10,11] Mark A Little,[12] Raashid A Luqmani,[13] Alfred Mahr,[14] Peter A Merkel ,[15] Aladdin J Mohammad ,[11,16] Sara Monti ,[17,18] Chetan B Mukhtyar ,[19] Jacek Musial,[20] Fiona Price-Kuehne,[11] Mårten Segelmark,[21] Y K Onno Teng ,[22] Benjamin Terrier ,[23] Gunnar Tomasson ,[24,25] Augusto Vaglio ,[26] Dimitrios Vassilopoulos ,[27] Peter Verhoeven,[28] David Jayne [11]

Hellmich B, *et al*. *Ann Rheum Dis* 2024;**83**:30–47. doi:10.1136/ard-2022-223764

Table 2 Examples of organ/life-threatening and not organ/life-threatening manifestations in patients with AAV

Examples of potentially organ/life-threatening manifestations*	Examples of manifestations that are not ultimately organ/life-threatening*
Glomerulonephritis	Nasal and paranasal disease without bony involvement (erosion) or cartilage collapse or olfactory dysfunction or deafness
Pulmonary haemorrhage	Skin involvement without ulceration
Meningeal involvement	Myositis (skeletal muscle only)
Central nervous system involvement	Non-cavitating pulmonary nodules
Retro-orbital disease	Episcleritis
Cardiac involvement	
Mesenteric involvement	
Mononeuritis multiplex	



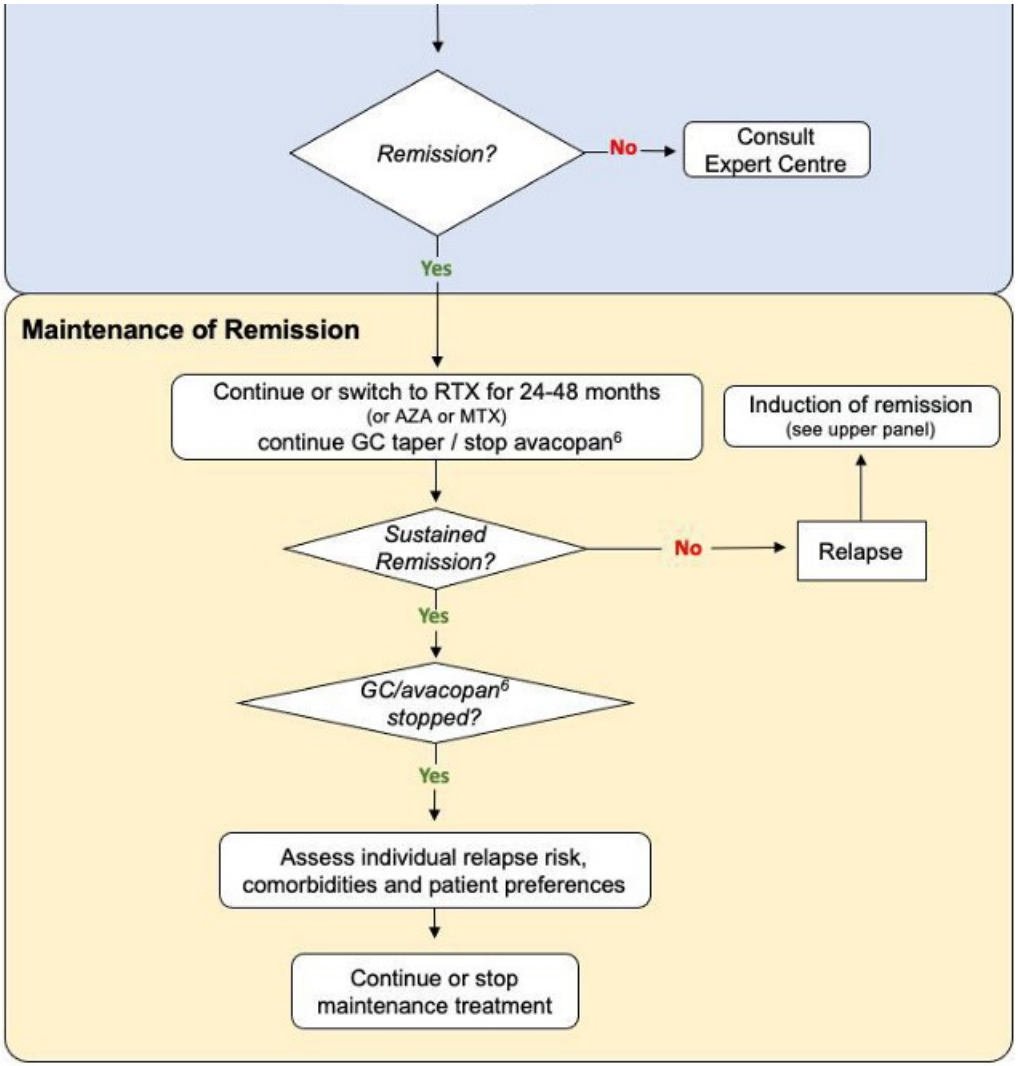
As part of regimens for induction of remission in GPA or MPA, we recommend treatment with oral glucocorticoids at a starting dose of 50–75 mg prednisolone equivalent/day, depending on body weight. We recommend stepwise reduction in glucocorticoids according to table 4 and achieving a dose of 5 mg prednisolone equivalent per day by 4–5 months.

Plasma exchange may be considered as part of therapy to induce remission in GPA or MPA for those with a serum creatinine >300 µmol/L due to active glomerulonephritis.*

Routine use of plasma exchange to treat alveolar haemorrhage in GPA and MPA is not recommended.†

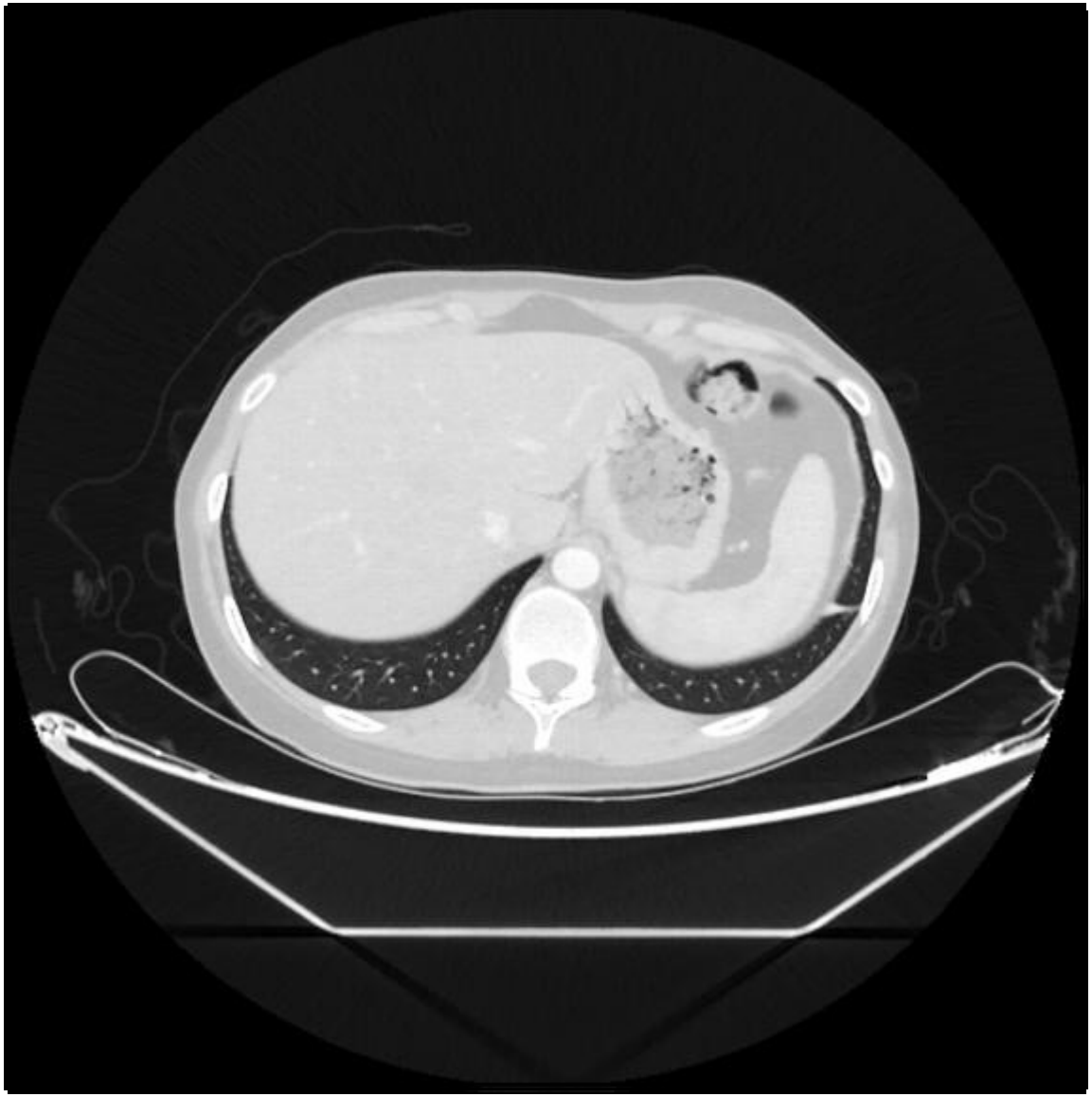
At 6th month of induction treatment

- No symptom
- PE: No pathological sign
- Lab: Hb: 13 gr/dl, Hct: %35
- Urine analysis: N
- Treatment with Rituximab every 6 months planned



We recommend that therapy to maintain remission for GPA and MPA be continued for 24–48 months following induction of remission of new-onset disease.* Longer duration of therapy should be considered in relapsing patients or those with an increased risk of relapse, but should be balanced against patient preferences and risks of continuing immunosuppression.†

14.06.2021



T.C
FIRAT ÜNİVERSİTESİ HASTANESİ Oluşturulma Tarihi : 11.01.2024 08:31:0

EPİKRİZ

Hasta Dosya No	: 809719	Başvuru No	:
Adı Soyadı	:	Telefon	: 05317191792
T.c Kimlik No	:	Yatış Tarihi	: 10.01.2024 13:1
Doğum Yeri	: ELAZIĞ	Cıkış/Tab. Tarih	: 11.01.2024 08:31 / 11.01.2024 00
Doğum Tarihi	: 03.05.1990	Bölüm	: GÖZ HASTALIKLARI KLİNİĞİ
Başvuru Tarihi	: 03.01.2024 09:25:00	Sorumlu Oğr.Uyesi	:
Kurumu	: SSK SAĞLIK İŞLERİ MÜDÜRLÜĞÜ	Sorumlu Başvurulan Dr:	
		Cıkış Sekli	: Haliyle Taburcu
Adres	: CUMHURİYET MAH. MAH. ŞEHİT KORGENERAL HULUSİ SAYIN CAD. SOK. 131/7 MERKEZ ELAZIĞ		

TANI

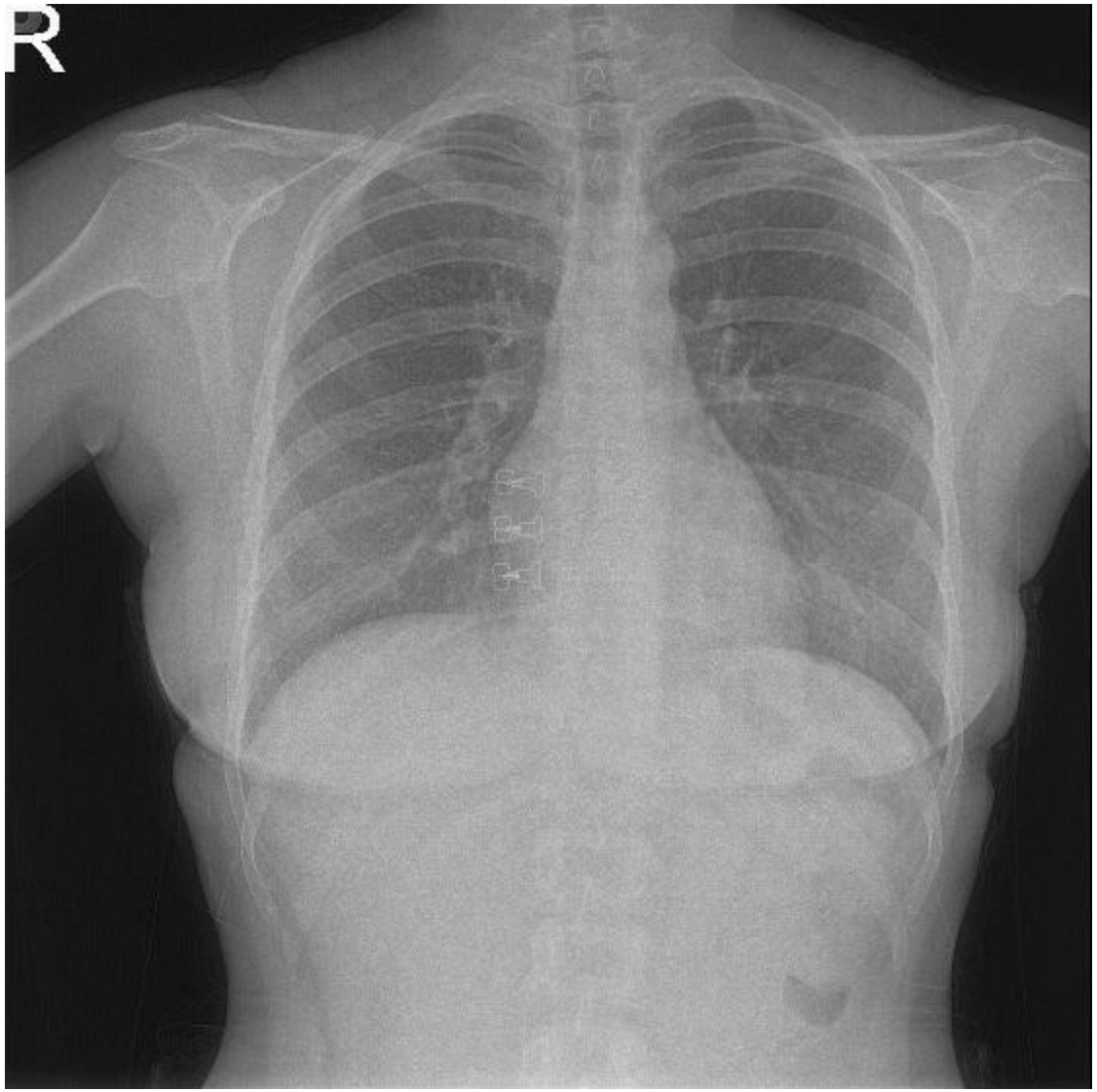
H26.0- İNFANTİL, JUVENİL VE PRESENİL KATARAKT Ana Tanı,H10.2- AKUT KONJONKTİVİTLER, DİĞER Ana Tanı,

KLİNİK SEYİR - TEDAVİ

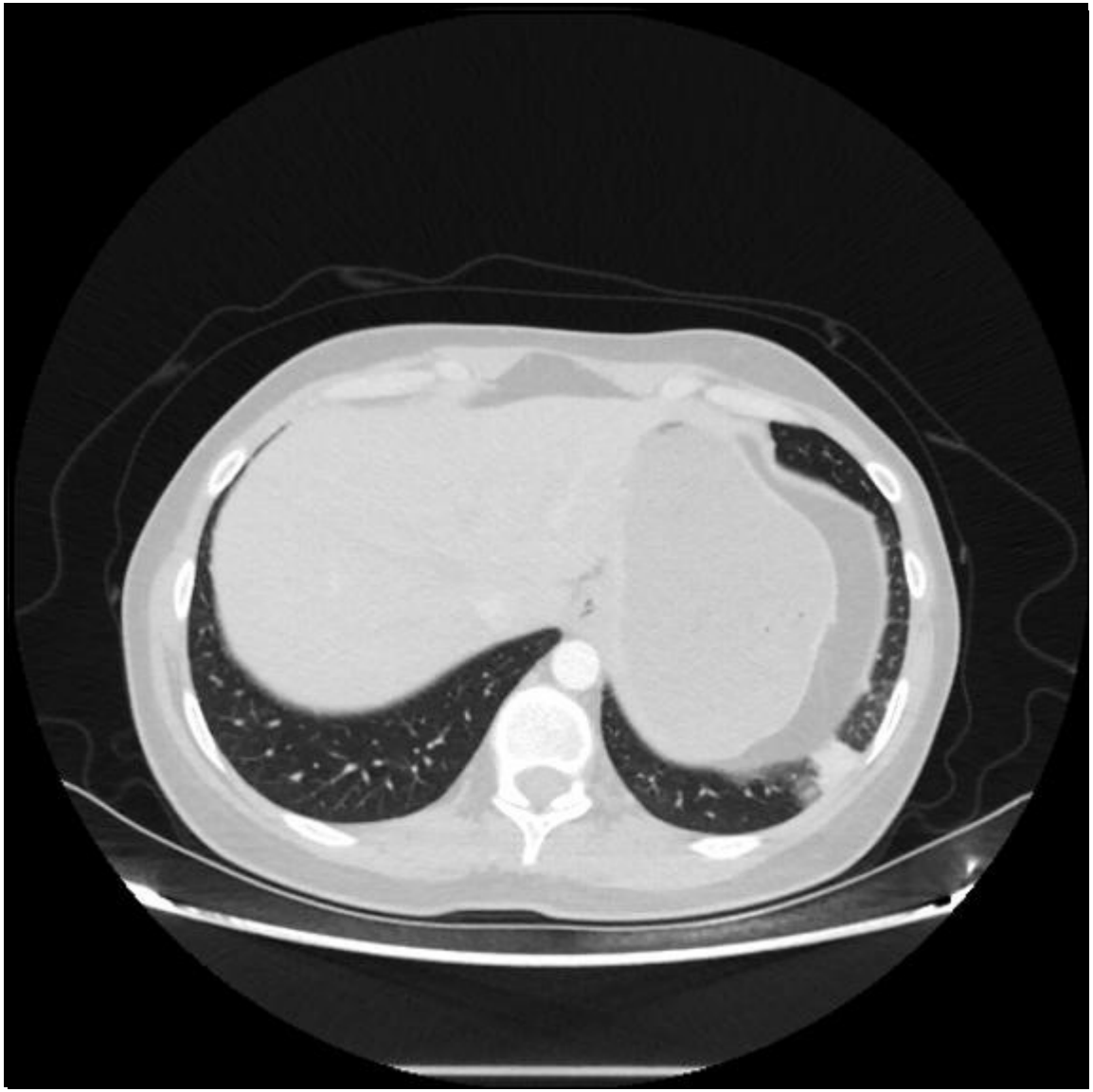
Sağ göze katarakt operasyonu önerilerek hasta kliniğimize yatırıldı.Hastadan hepatit markırları istendi. Rejyonel oküler anestezi altında sağ göz FAKO+İOL implantasyonu operasyonu yapıldı. Hastaya moksifloksasin damla 5x1, deksametazon damla 5x1 tedavisi başlandı. Klinik takiplerinde genel durumu stabil olan hasta moksifloksasin damla 5x1, deksametazon damla 5x1 tedavisi ile 1 gün ve 1 hafta sonra poliklinik kontrolüne gelmek üzere taburcu edildi

After 3 years she applied to the eye disease clinic, operated because of cataract

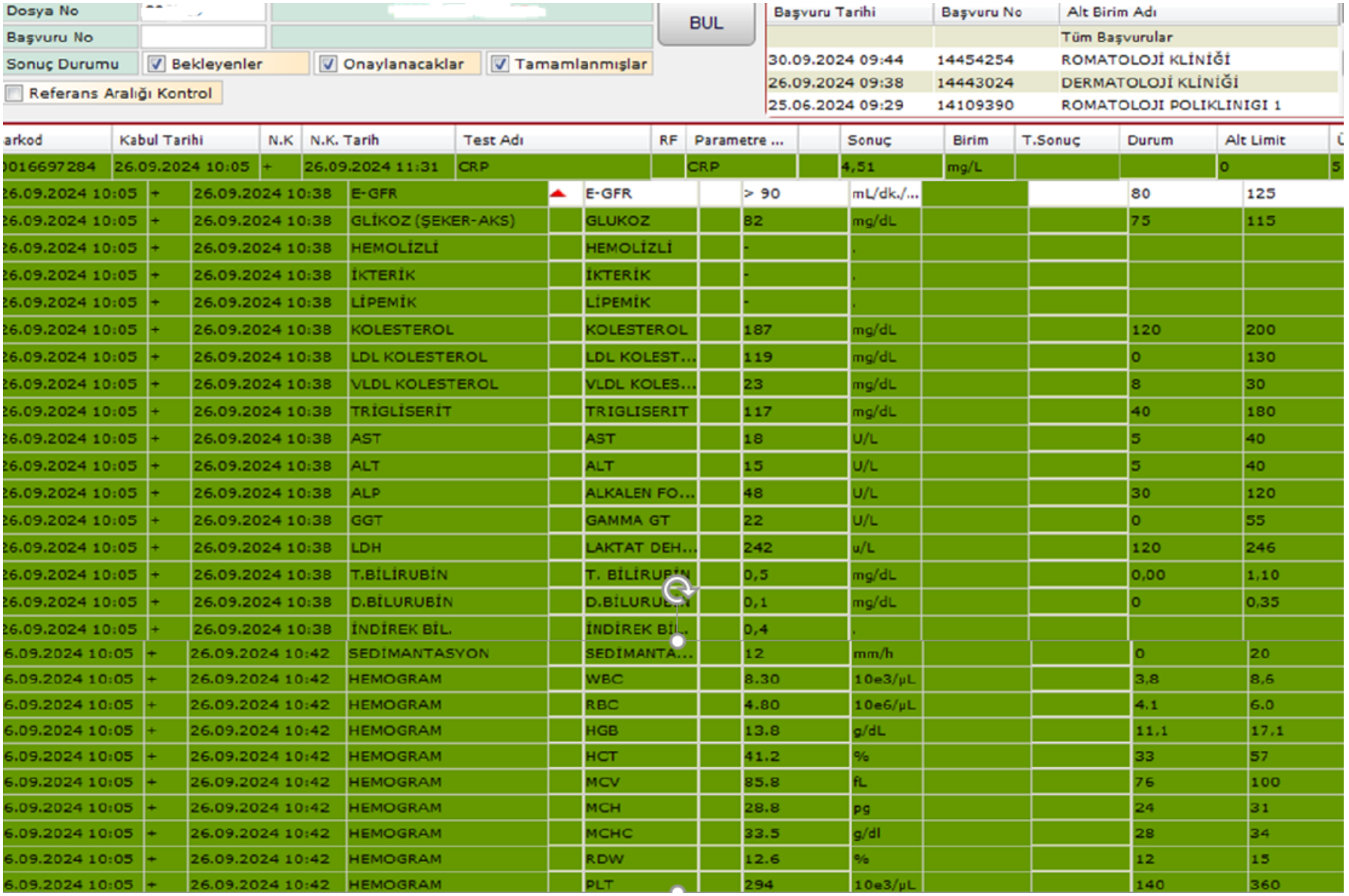
06.02.2024



06.02.2024



Last visit: 26.09.2024



Dosya No
Başvuru No
Sonuç Durumu ☑ Bekleyenler ☑ Onaylanacaklar ☑ Tamamlanmışlar
☐ Referans Aralığı Kontrol
BUL

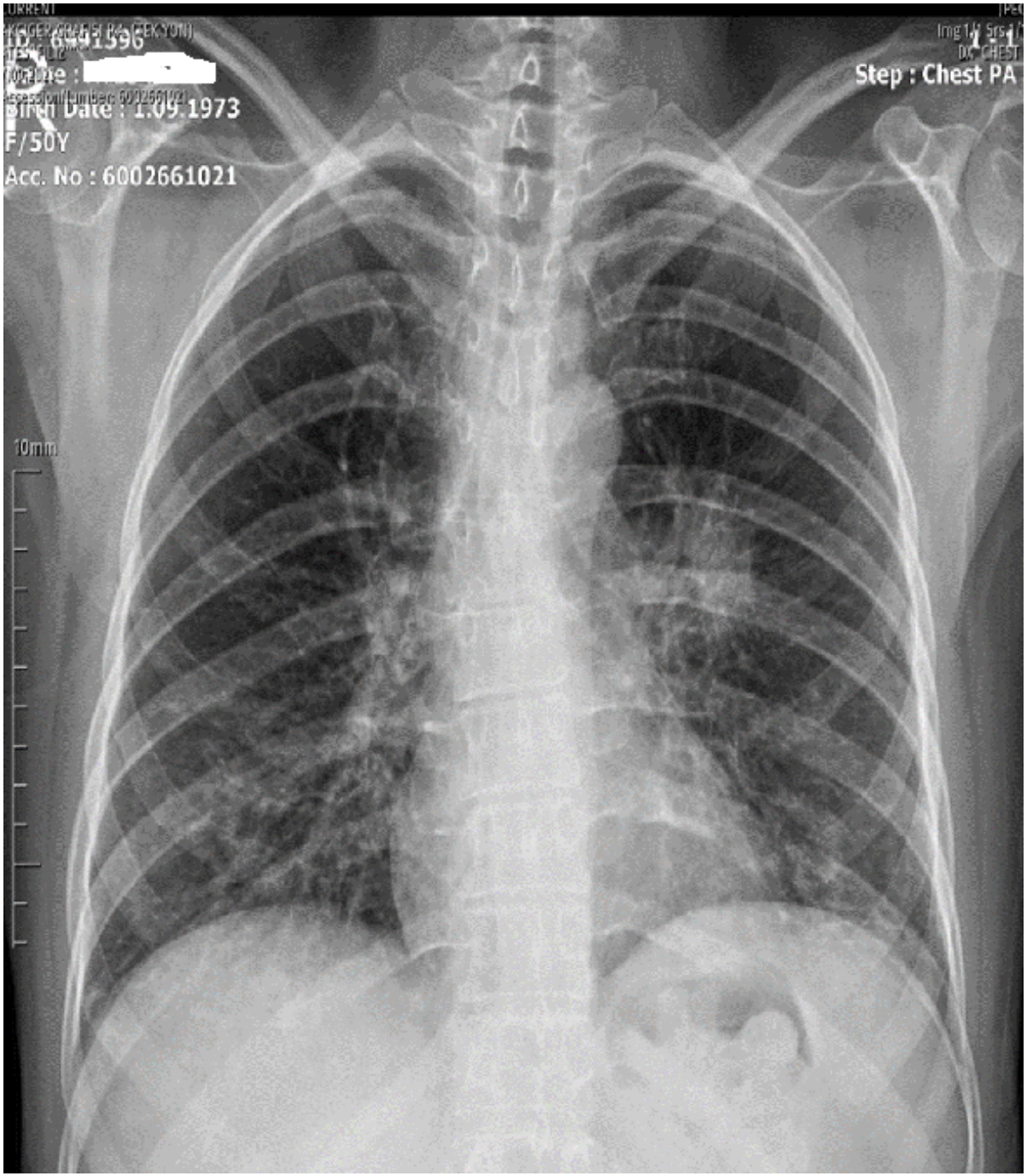
Başvuru Tarihi	Başvuru No	Alt Birim Adı
		Tüm Başvurular
30.09.2024 09:44	14454254	ROMATOLOJİ KLİNİĞİ
26.09.2024 09:38	14443024	DERMATOLOJİ KLİNİĞİ
25.06.2024 09:29	14109390	ROMATOLOJI POLIKLINIGI 1

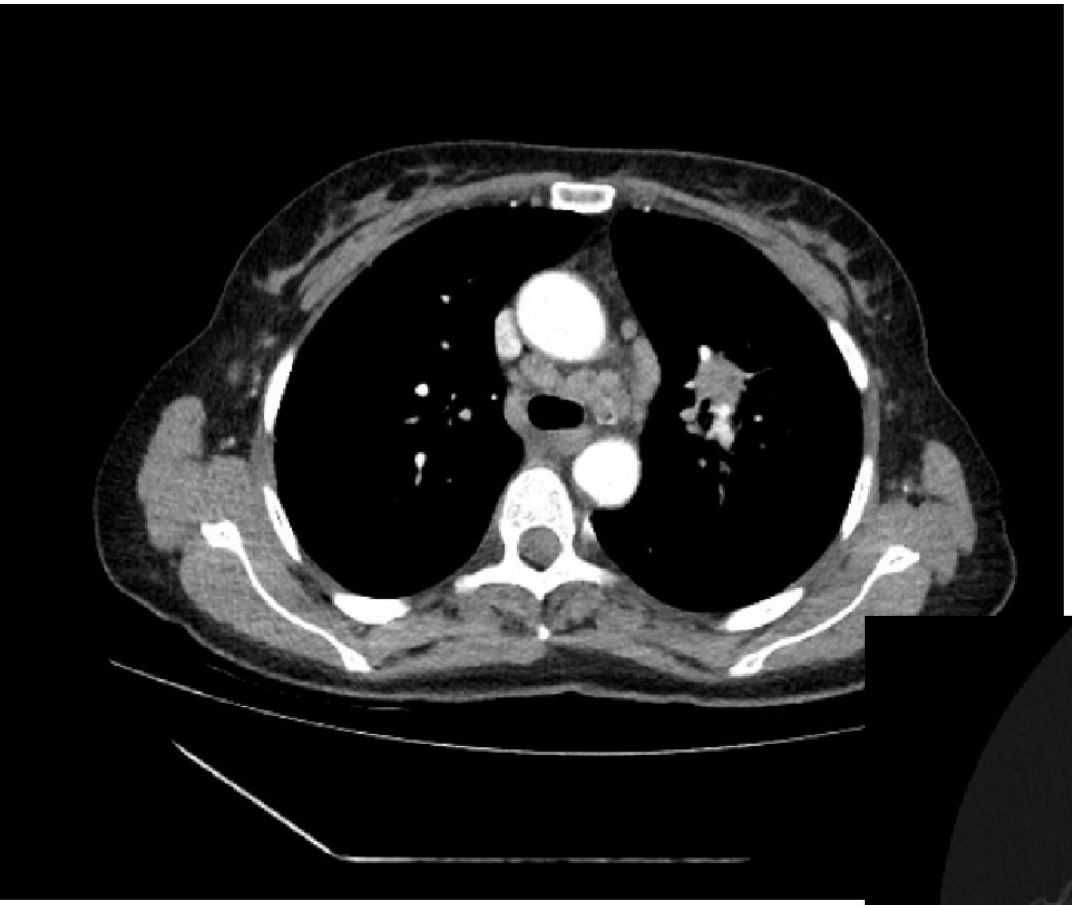
arkod	Kabul Tarihi	N.K	N.K. Tarih	Test Adı	RF	Parametre ...		Sonuç	Birim	T.Sonuç	Durum	Alt Limit	Ü
0016697284	26.09.2024 10:05	+	26.09.2024 11:31	CRP		CRP		4,51	mg/L			0	5
26.09.2024 10:05	+	26.09.2024 10:38	E-GFR		E-GFR		> 90	mL/dk./...			80	125	
26.09.2024 10:05	+	26.09.2024 10:38	GLİKOZ (ŞEKER-AKS)		GLUKOZ		82	mg/dL			75	115	
26.09.2024 10:05	+	26.09.2024 10:38	HEMOLİZLİ		HEMOLİZLİ		-	,					
26.09.2024 10:05	+	26.09.2024 10:38	İKTERİK		İKTERİK		-	,					
26.09.2024 10:05	+	26.09.2024 10:38	LİPEMİK		LİPEMİK		-	,					
26.09.2024 10:05	+	26.09.2024 10:38	KOLESTEROL		KOLESTEROL		187	mg/dL			120	200	
26.09.2024 10:05	+	26.09.2024 10:38	LDL KOLESTEROL		LDL KOLEST...		119	mg/dL			0	130	
26.09.2024 10:05	+	26.09.2024 10:38	VLDL KOLESTEROL		VLDL KOLES...		23	mg/dL			8	30	
26.09.2024 10:05	+	26.09.2024 10:38	TRİGLİSERİT		TRIGLISERIT		117	mg/dL			40	180	
26.09.2024 10:05	+	26.09.2024 10:38	AST		AST		18	U/L			5	40	
26.09.2024 10:05	+	26.09.2024 10:38	ALT		ALT		15	U/L			5	40	
26.09.2024 10:05	+	26.09.2024 10:38	ALP		ALKALEN FO...		48	U/L			30	120	
26.09.2024 10:05	+	26.09.2024 10:38	GGT		GAMMA GT		22	U/L			0	55	
26.09.2024 10:05	+	26.09.2024 10:38	LDH		LAKTAT DEH...		242	u/L			120	246	
26.09.2024 10:05	+	26.09.2024 10:38	T.BİLİRUBİN		T. BİLİRUBİN		0,5	mg/dL			0,00	1,10	
26.09.2024 10:05	+	26.09.2024 10:38	D.BİLURUBİN		D.BİLURUBİN		0,1	mg/dL			0	0,35	
26.09.2024 10:05	+	26.09.2024 10:38	İNDİREK BİL.		İNDİREK BİL.		0,4	,					
26.09.2024 10:05	+	26.09.2024 10:42	SEDİMANTASYON		SEDİMANTA...		12	mm/h			0	20	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		WBC		8.30	10e3/µL			3,8	8,6	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		RBC		4.80	10e6/µL			4.1	6.0	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		HGB		13.8	g/dL			11,1	17,1	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		HCT		41.2	%			33	57	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		MCV		85.8	fL			76	100	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		MCH		28.8	pg			24	31	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		MCHC		33.5	g/dl			28	34	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		RDW		12.6	%			12	15	
26.09.2024 10:05	+	26.09.2024 10:42	HEMOGRAM		PLT		294	10e3/µL			140	360	

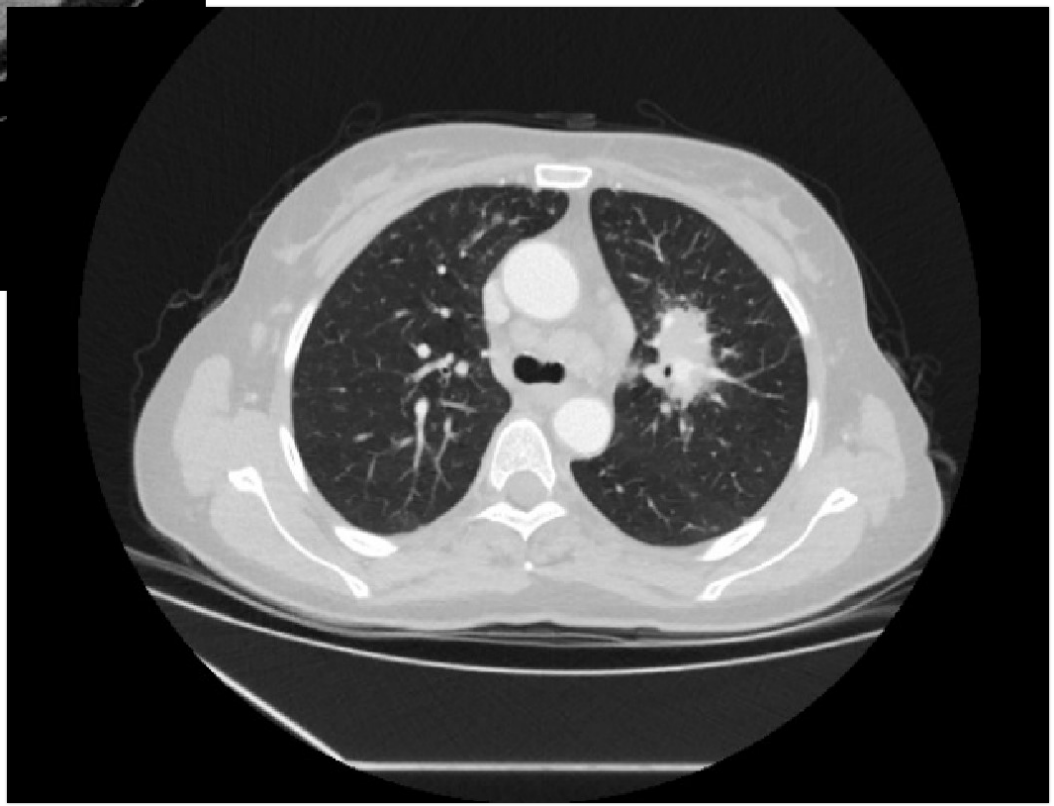
30.09.2024 10:05	+	30.09.2024 11:14	İDRAR KREATİNİN (SP...		İDRAR KRE...		20.09	mg/dL				
30.09.2024 10:05	+	30.09.2024 11:14	İDRAR PROTEİNİ(SPOT)		İDRAR PRO...	R	6	mg/dL	Error		1	14
30.09.2024 10:05	+	30.09.2024 11:14	SPT.İDR PRO/SPT.İDR ...	▲	SPT.İDR PR...		0,3	mg/dL			0,04	0,06
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ	▲	Dansite		1022	.			<1.030	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		PH		6	.			6,0-8,0	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Protein		Neg	mg/dL			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Glukoz		Neg	mg/dL			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Keton		Neg	mmol/L			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Nitrite		Neg	.			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Bilirubin		Neg	mg/dL			NEGATIF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Urobilinojen		Normal	mg/dL			NORMAL	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Eritrosit		+-	Ery/uL			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Renk		Light Yellow	.			SARI	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Lökosit Este...		+-	Leu/uL			NEGATİF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Lökosit Kü...		0					
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Lökosit		12	HPF			<5/HPF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Eritrosit		10	HPF			<3/HPF	
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Eritrosit Kü...		0					
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Bakteri		0	HPF				
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Kalsiyum O...		0	HPF				
26.09.2024 10:05	+	26.09.2024 11:09	TAM İDRAR TETKİKİ		Hyalin Silen...		0	HPF				

Case-3

- 50 y, F
- Symptoms: Cough, phlegm
- IVIG for Common Variable Immunodeficiency diagnosis (30 gr/month)
- Allergic rhinitis (+), Asthma (+)


Birth Date : 1.09.1973
F/50Y
Acc. No : 6002661021
Step : Chest PA
10mm





- Sputum ARB 3 times (-), no growth in TB culture

FOB

- Lavage ARB (-), no growth in TB culture
- BAL: % 11 lymphocyte

  % 54 macrophage

  % 35 neutrophil

  CD4/CD8: 1.7

  - Cytology: Benign inflammatory BAL finding with **significant neutrophil** leukocyte increase
  - Tru-cut bx: **Fibrosis and inflammation**

- **EBUS:** 4R, 7, 11L, 11R lymph nodes were sampled
- TBB applied

- Diagnosis: **Granulomatous inflammation**

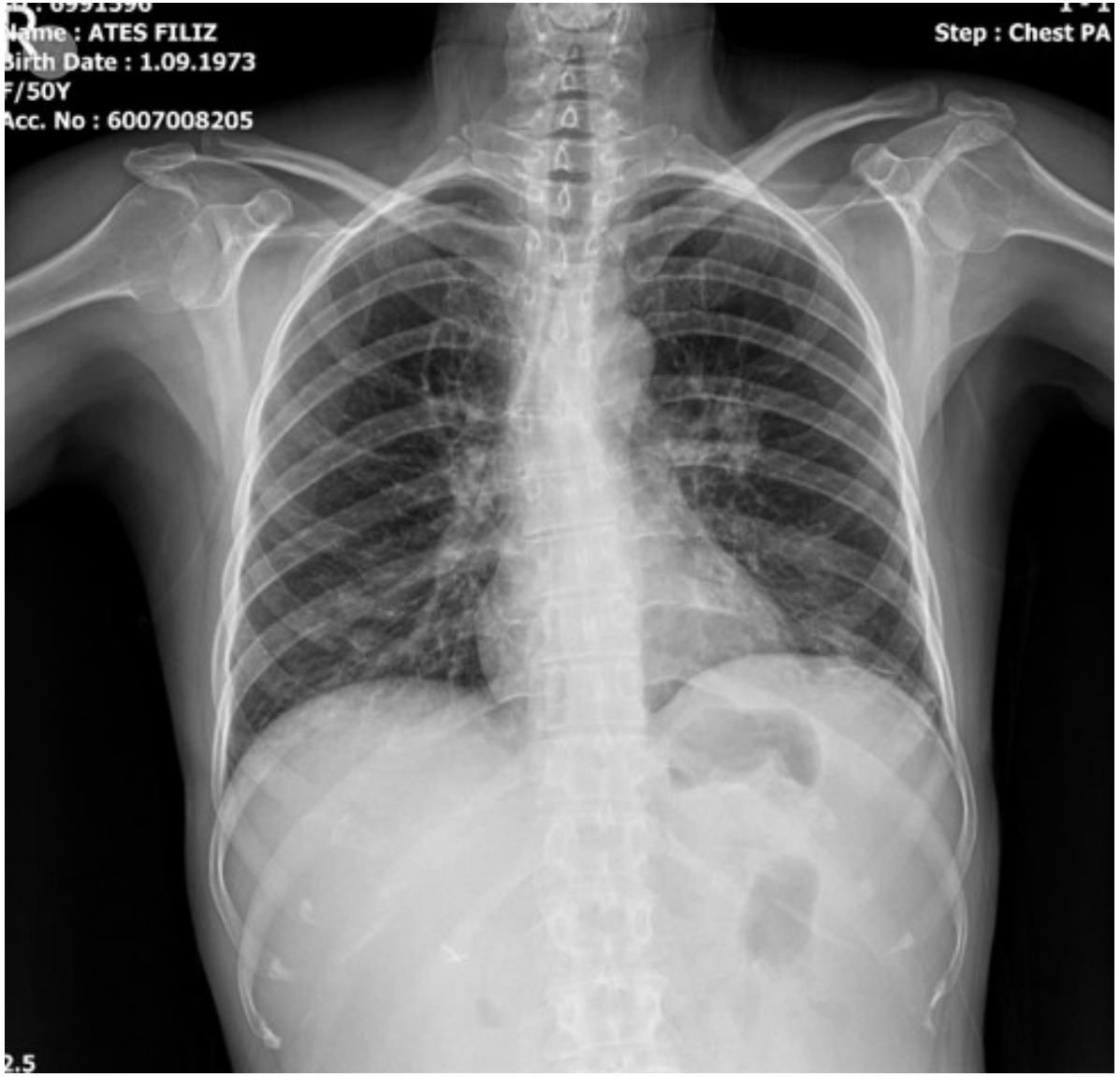
TANI

GRANÜLOMATÖZ İNFLAMASYON BULGULARI, BRONŞ BİYOPSİSİ.

YORUM

Olgunun, sarkoidozis ve tbc başta olmak üzere olası granülomatöz inflamasyon etyolojileri yönünden araştırılması öneriliı

- Lavage ARB (-)
- TB PCR (-)
- No growth in TBC culture
- TST: 4 mm
- IGRA: (-)
- Patient was diagnosed as **GLILD** because of CVID
- IVIG and streoids prescribed


ame : ATES FILIZ
irth Date : 1.09.1973
F/50Y
Acc. No : 6007008205
Step : Chest PA
2.5

Granulomatous and Lymphocytic Interstitial Lung Disease

- GLILD is a severe non-infectious complication of CVID
- GLILD develops in 8-20% of CVID patients
- GLILD is a significant cause of morbidity and premature mortality among patients with CVID
- GLILD has been defined as "a distinct clinico-radio-pathological ILD occurring in patients with CVID, associated with a lymphocytic infiltrate and/or granuloma in the lung, and in whom other conditions have been considered and where possible excluded"

Hurst JR, et al. British lung foundation/United Kingdom primary immunodeficiency Network consensus statement on the definition, diagnosis, and management of granulomatous-lymphocytic interstitial lung disease in common variable immunodeficiency disorders. J Allergy Clin Immunol Pract. 2017;5(4):938–945

Predictors for GLILD diagnosis

- Splenomegaly
- Autoimmune cytopenias
- Low IgG, IgM and IgA levels
- Increased CD21lo B cells percentage
- Lower percentage of switched-memory B cells and marginal zone B cells
- Low TLC, FVC, and DLCO %pred

Clinical manifestation

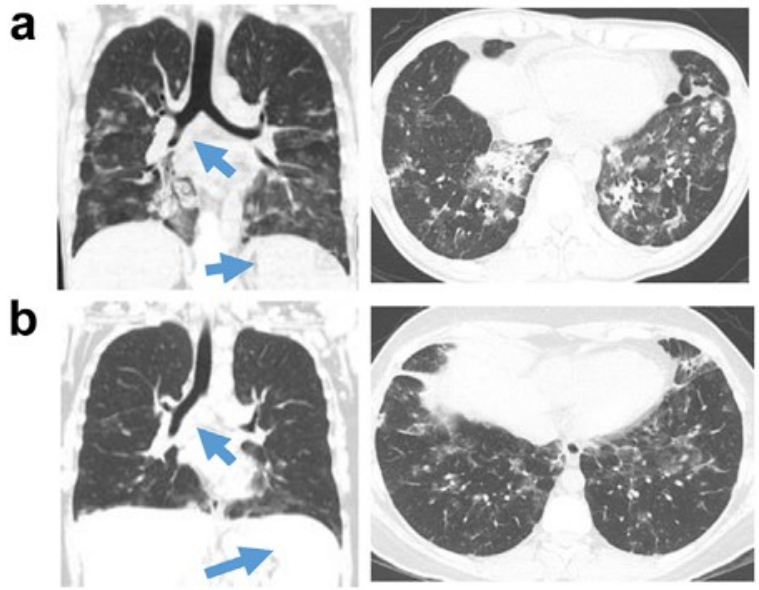
- 20-50 years
- Higher prevalence among females
- 15% of patients may be asymptomatic
- Symptoms; exertional dyspnea and nonproductive cough
- Production of sputum, wheezing can be seen in patients with concomitant bronchiectasis
- PE: Inspiratory crackles, wheezing, splenomegaly, lymphadenopathy

Approach to diagnosing and managing granulomatous-lymphocytic interstitial lung disease

eClinicalMedicine
2024;75: 102749

Jessica Galant-Swafford,[a] Jason Catanzaro,[b] Rosane Duarte Achcar,[c] Carlyne Cool,[d] Tilman Koelsch,[e] Tami J. Bang,[e] David A. Lynch,[e] Rafeul Alam,[a] Rohit K. Katial,[a] and Evans R. Fernández Pérez[f,*]

HRCT Pattern[a]	
Typical GLILD	Distribution – Axial: peribronchovascular predominance – Craniocaudal: lower lung zones Features – All of the following features: nodularity, ground-glass opacity, consolidation Other findings – Hilar, mediastinal lymphadenopathy, splenomegaly



Approach to diagnosing and managing granulomatous-lymphocytic interstitial lung disease

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Histopathological Pattern[b]	
Typical GLILD	Major features – Presence of two major features in at least one of the sampled lobe(s) of the lung (surgical lung biopsy): 1. Pulmonary lymphoid hyperplasia with reactive germinal centers in the form of: – Follicular bronchiolitis, And/or – Nodular lymphoid hyperplasia, And/or – Any interstitial lymphocytic infiltrates 2. Granulomas (well, moderately, or poorly-formed) Secondary features 1. Organizing pneumonia 2. Interstitial scarring – Lack of features of an alternative diagnosis

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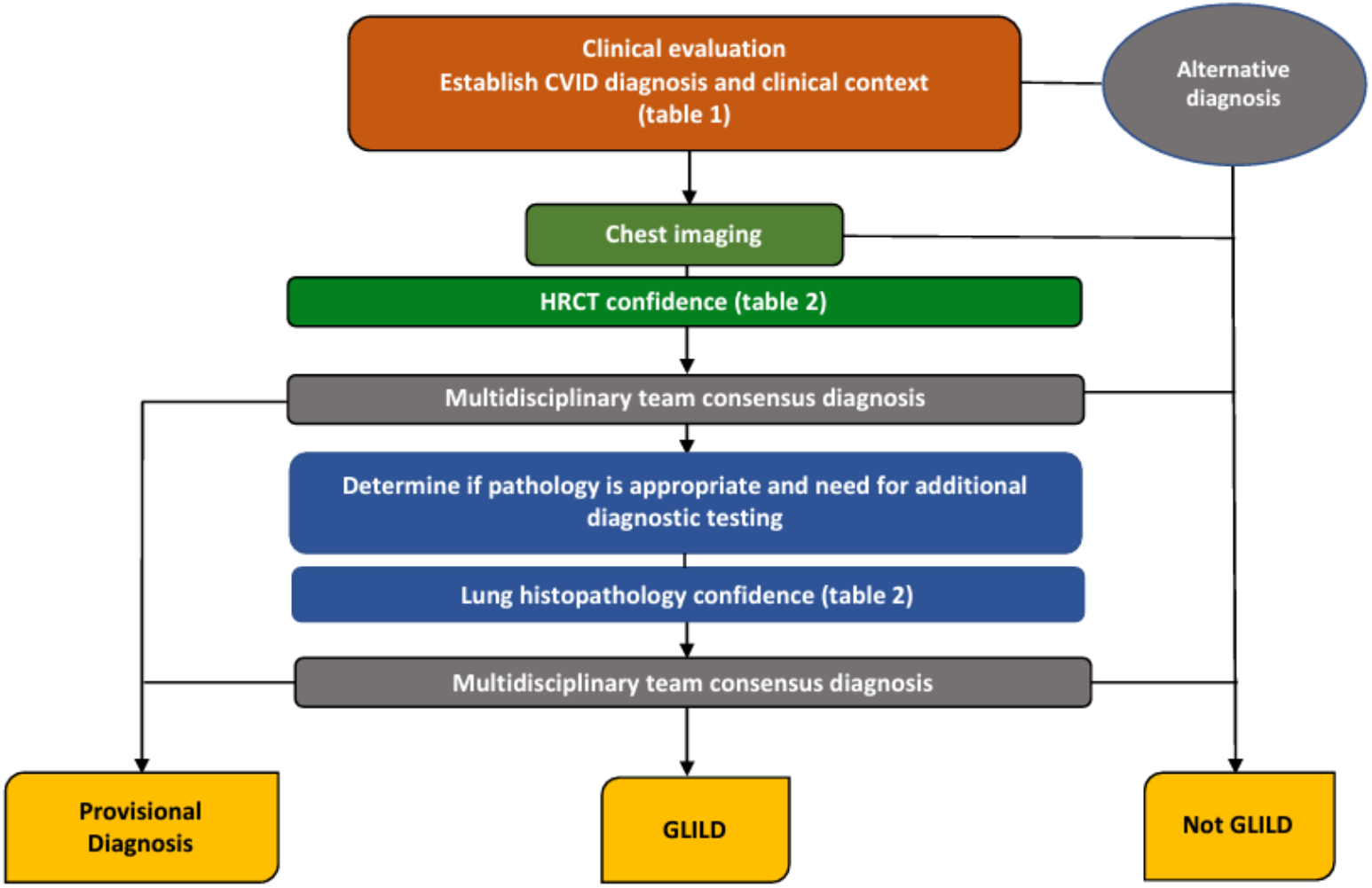

Table 1. Differences in Granulomatous lymphocytic interstitial lung disease and sarcoidosis

Feature	GLILD	Sarcoidosis
Organ system involvement (22, 55)		
Pulmonary	51%	95%
Spleen	46%	6.7
Lymph node		15.2
Liver	41%	11.5
Skin	7%	15.9
Bone marrow	8%	3,9%
CNS	5%	4.6%
GI tract	15%	Rare
Recurrent infections	Common	May occur if architectural distortion of lung
Autoimmunity	Frequently report	Not seen
Immunoglobulin levels	Low	Normal or high. May be low in patients on long term steroids
Chest CT		
Distribution	Could have lower lobe disease	Upper lobe predominant disease
Common finding	Larger nodule with random or perilymphatic distribution	Perilymphatic micronodular infiltrate in bronchovascular distribution
Flame shape hemorrhage, 'halo" sign (16)	More common than sarcoidosis	Could be seen
Bronchiectasis	Common due to recurrent infections	Cicatricial bronchiectasis in setting of architectural distortion
Mediastinal / hilar adenopathy	Present	More prominent
Bronchoalveolar fluid findings		
Cultures	Rules out infection in cases of CVID (42)	Usually negative. Must rule our etiologies that may mimic clinical or radiologic manifestations of sarcoidosis such as histoplasma or tuberculosis
CD4: CD8 ratio	Usually, normal	High (> 3.5)

Treatment

- IGRT should be optimized before the initiation of immunosuppressive therapy
- Immunosuppressive treatment (steroid, MMF, AZA), rituximab
- Hematopoietic stem cell transplantation (HSCT) is a potential definitive therapy

Thank you for your attention...