ON-LINE SUPPLEMENTARY MATERIAL FOR NGT PROCEEDINGS

This supplement expands on the details and voting results of the NGT process. The voting items and results reflect end-products of an iterative process that took place over three years. The Delphi process enlisted the wide participation of the ILD medical expert community of pulmonary and rheumatology specialists, with support from an advisory panel of pathologists and radiologists to identify domains and produce a list of instruments with which to measure these domains that are acceptable to the greater community of ILD experts. **Patient Participation**: In order to proceed with the stages beyond the Delphi, patient perspective of ILD was factored into the results of the Delphi process based on focus groups with 45 patients with CTD-ILD and the results of a prior study with 20 patients with IPF (conducted by: Swigris JJ, et al. Patients' perspectives on how idiopathic pulmonary fibrosis affects the quality of their lives. Health Qual Life Outcomes. 2005).

I. NGT Participation:

Voting at the NGT was done through a pre-programmed automated response system which collected voting responses onto a computer hard drive and grouped according to participant type. Fourteen pulmonary specialists, 16 rheumatology specialists, 2 radiologists and 6 CTD-ILD patient partners and 2 IPF patient partners were invited to participate with following participants in actual attendance (of which 10 pulmonary, 12 rheumatology and 1 radiology specialist participated with ultimately a patient representing each IPF, IIM-ILD, RA-ILD and SSc-ILD participating):

Patient Research Partners: Diseases represented were IPF, RA-ILD, IIM-ILD and SSc-ILD. Several patients had clinical trial experience. All patients had the experience of oxygen dependency. Three had disease severe enough to have either received or are being considered for transplantation.

Robert Hedlund
Catherine Sarver
Karen Nichols
Pieter van den Assum
Daphne LeSage (involved in the development of the NGT proceedings but unable to attend due to inclement weather resulting in airport closure)

Pulmonary ILD Specialists:

Katerina Antoniou
Robert P. Baughman
Kevin K. Brown
Kevin Flaherty
Kristin B. Highland (Trained in Rheumatology)
Dong Soon Kim
Luca Richeldi
Jay H. Ryu
Jeffrey Swigris
Athol Wells

Rheumatology Specialists:

Paul F. Dellaripa
Oliver Distler
Aryeh Fischer
Dinesh Khanna
Eric L. Matteson
Peter A. Merkel
Frederick W. Miller
Shikha Mittoo
Chester V. Oddis
Susanna Proudman
James R. Seibold
Vibeke Strand

Radiology ILD Specialist:

David Lynch (Trained in internal medicine and radiology, his votes were attributed to the Pulmonary Specialty Group, thus tabulated for both IPF and CTD-ILD)

Convener/Organiser/Methods Supervisor/Patient Educational Sessions: Lesley Ann Saketkoo **Moderation**: Peter A. Merkel, Oliver Distler

II. Domain Teams:

DYSPNEA:
Robert P. Baughman
Kevin Flaherty
Dinesh Khanna
Catherine Sarver

COUGH:
Robert P. Baughman
Shikha Mittoo
Daphne LeSage
Jeffrey Swigris

HRQoL:
Aryeh Fischer
Kevin Flaherty
Dinesh Khanna
Peter A. Merkel
Karen Nichols
Susanna Proudman
Vibeke Strand
Jeffrey Swigris

LUNG PHYSIOLOGY:
Kevin Flaherty
Kristin B. Highland
Dong Soon Kim
Otylia Kowal-Bielecka
Frederick W. Miller
Susanna Proudman
Jay H. Ryu

LUNG IMAGING:
Katerina Antoniou
Oliver Distler
David Lynch
Jay H. Ryu
Athol U. Wells

SURVIVAL:
Kevin K. Brown
Paul F. Dellaripa
Robert Hedlund
Eric L. Matteson
Athol U. Wells

MEDICATIONS:
Chester V. Oddis
James R. Seibold
Vibeke Strand

DOMAIN TEAMS Oversight: Lesley Ann Saketkoo

III. OMERACT (Outcome Measures in Rheumatology [initially 'for Clinical Trials' though this is no longer part of the official title])

Background: OMERACT is an international non-profit organization established in 1992 dedicated to the identification and development of appropriate outcome measures in disease. OMERACT provides a home for many disease-based working groups investigating outcome measures for use in clinical trials. OMERACT has characterized validity in terms of a 'filter' that provides an organizational checklist for an instrument's ability to satisfy accepted components of validity.

Filter: The components of the filter are grouped under three main criteria: truth, discrimination and feasibility. While the ideal instrument would satisfy all three criteria completely, it is recognized that many useful instruments do not. The filter serves as a guide to identifying the degree to which instruments have demonstrated validity.

Glossary of Terms/Properties comprised in the OMERACT Filter:

Truth:

Face Validity: The instrument hypothetically or at 'face value' makes sense; usually brings the instrument into consideration for study.

Content Validity: The instrument has demonstrated ability to measure the intended concept/domain; i.e. the substance of a measure is acknowledged to reflect a concept /domain well in regards to relevant content and comprehension for a specific disease. This may be gleaned from prior studies or active presentation to and/or item collection from medical experts and/or patients.

Construct Validity: The instrument has been applied in real world setting and demonstrates confirmatory relationships with other accepted measures for that disease – whether the relations are convergent (correlative when anticipated to be so) or divergent (non-correlative when anticipated not to be so) with other accepted outcome measures in that disease. This operational step provides confirmation for further investigation of the instrument.

Criterion Validity: The values provided by the instrument correlate with or predict results of the accepted 'gold standard' for that disease.

Discrimination:

Discrimination: The instrument has demonstrated ability to be responsive to changes for the intended concept/domain; while remaining sufficiently unresponsive to other like or confounding situations.

Reliability: The instrument demonstrates reproducibility and, importantly, accurate values over multiple measurements.

Sensitivity to Change: The values of the instrument demonstrate incremental results that either positively or negatively correlate with changes of the disease over time; e.g. while an instrument may have tremendous diagnostic value, it may not be useful to monitor the course of a disease.

Feasibility: Focuses on logistical and practical implementation and is often the deciding factor on the utility of an instrument.

Interpretability: Analysis/computation of results is sufficiently straightforward and undemanding so as not to introduce potential errors or hardship in implementation or interpretation of results.

Accessibility: There is little or no impediment to the instrument being commonly (or potentially) available for use; and the financial costs and time burden of obtaining, implementation and interpretation of the instrument does not impose unusual hardship.

Safety: The instrument poses little or no risk to patients or personnel implementing the measure.

IV. Post-Delphi Introduction of Items

Domain or Instrument Introduced	**Support for Post-Delphi Introduction**
Domain of *Cough*	Substantiated by Patient Perspective in both CTD-ILD and IPF (Swigris et al).
Instrument of Leicester Cough Questionnaire (LCQ) as a measure of *Cough*	Identified by comparative analysis by *Cough* domain team, discussion and Delphi voting as the most appropriate measure to supply an instrument under cough.
Instrument of Mahler Dysnea Index (MDI) as measure of *Dyspnea*	Identified by updated literature review and voted upon as having substantive findings warranting NGT discussion and voting. Exclusion of this item was collectively viewed as injurious to fair representation of post-Delphi evidence.
Instrument of University of California San Diego Shortness of Breath Questionnaire (UCSD-SBQ) as measure of *Dyspnea*	Identified by updated literature review and voted upon as having substantive findings warranting NGT discussion and voting. Exclusion of this item was collectively viewed as injurious to fair representation of post-Delphi evidence.
The concepts of *Fatigue*, *Participation*, *Physical Function*, *Self-care* and *Sleep*	These concepts were identified as important in Patient Perspective studies. It was agreed that disease-specific investigations into HRQoL would incorporate these components.
The measure of All Cause Mortality as a measure of *Survival*	Identified as an important generic identifier of death in clinical trials.

V. VOTING RESULTS:

For the following series of tables the purple shaded columns are the total responses of the groups appropriated to IPF or to CTD-ILD. While the pink shaded columns are the individual groups whose votes are appropriated to the total accepted votes. Acceptance was agreed upon a priori as ≥70% with the following tabulations: **IPF Voting**: Pulmonary Specialists + IPF Patient Partners

CTD-ILD Voting: Pulmonary Specialists + Rheumatology Specialists + CTD-ILD Patient Partners

Dyspnea IPF

Instrument		**Total for Acceptance**	Pulms	IPF Patient
Dyspnea 12		70% (7/10)	67% (6/9)	100% (1/1)
MRC		92% (11/12)	91% (10/11)	100% (1/1)
UCSD		80% (8/10)	78% (7/9)	100% (1/1)
Borg		36% (4/11)	40% (4/10)	0% (0/1)
Possible Secondary End-Point		82% (9/11)	80% (8/10)	100% (1/1)
Need New Patient Derived Instrument		73% (8/11)	70% (7/10)	100% (1/1)
Dyspnea 12 to be further evaluated	RESEARCH	100% 23/23	Show of Hands Voting from All Groups	

Dyspnea CTD-ILD

Instrument		**Total for Acceptance**	Pulms	Rheums	All Physicians	CTD ILD Patients
Dyspnea 12		88% (22/25)	80% (8/10)	92% (11/12)	86% (19/22)	100% (3/3)
MRC		75% (18/24)	78% (7/9)	75% (9/12)	76% (16/21)	66% (2/3)
Borg		32% (8/25)	30% (3/10)	33% (4/12)	32% (7/22)	33% (1/3)
MDI		58% (14/24)	40% (4/10)	67% (8/12)	55% (12/22)	100% (2/2)
MDI for SSc		54% (13/24)	50% (5/10)	55% (6/11)	52% (11/21)	66% (2/3)
Possible Secondary End-Point		96% (24/25)	90% (9/10)	100% (12/12)	95% (21/22)	100% (3/3)
Need New Patient Derived Instrument		76% (19/25)	70% (7/10)	92% (11/12)	82% (18/22)	33% (1/3)
MDI for future study	RESEARCH	91% 21/23	Show of Hands Voting from All Groups			

Cough in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
Leicester Cough Questionnaire as Interim Instrument		82% (9/11)	80% (8/10)	100% (1/1)
Possible Secondary End-Point		Agreement without dissension		
Need New Patient Derived Instrument		73% (8/11)	70% (7/10)	100% (1/1)

Cough in CTD ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD ILD Patients**
Leicester Cough Questionnaire as Interim Instrument		79% (19/24)	70% (7/10)	83% (10/12)	77% (17/22)	100% (2/2)
Possible Secondary End-Point		Agreement without dissension.				
Need New Patient Derived Instrument		64% (16/25)	60% (6/10)	75% (9/12)	68% (15/22)	33% (1/3)

Patient Global Assessment of Disease Activity in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
Pt-GA		64% (7/11)	60% (6/10)	100% (1/1)
Possible Secondary End-Point		90% (9/10)	89% (8/9)	100% (1/1)
10mm Change is Clinically Meaningful		30% (3/10)	22% (2/9)	100% (1/1)
PtGA further evaluated as Outcome Measure	RESEARCH	100% 23/23	Show of Hands Voting from all Groups	

Patient Global Assessment of Disease Activity in CTD ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD ILD Patients**
Pt-GA		96% (23/24)	100% (10/10)	92% (11/12)	95% (21/22)	100% (2/2)
Possible Secondary End-Point		92% (23/25)	80% (8/10)	100% (12/12)	91% (20/22)	100% (3/3)
10mm Change is Clinically Meaningful		71% (17/24)	50% (5/10)	83% (10/12)	68% (15/22)	100 (2/2)

Health Related Quality of Life in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
SF-36		82% (9/11)	80% (8/10)	100% (1/1)
SGRQ		82% (9/11)	80% (8/10)	100% (1/1)
Possible Secondary End-Point		100% (11/11)	100% (10/10)	100% (1/1)
Need New Patient Derived Instrument		90% (9/10)	90% (9/10)	Not Voted

Health Related Quality of Life in CTD ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD ILD Patients**
SF-36		100% (24/24)	100% (10/10)	100% (11/11)	100% (21/21)	100% (3/3)
SGRQ		87% (20/23)	90% (9/10)	82% (9/11)	86% (18/21)	100% (2/2)
HAQ-DI		54% (13/24)	30% (3/10)	64% (7/11)	48% (10/21)	100% (3/3)
Possible Secondary End-Point		100% (24/24)	100% 10/10	100% (11/11)	100% (21/21)	100% (3/3)
Need New Patient Derived Instrument		100% (22/22)	100% (8/8)	100% (11/11)	100% (19/19)	100% (3/3)

Lung Imaging: During the NGT, it was proposed by the Lung Imaging Team and agreed upon by the assembled group, that overall extent of disease in IPF is fibrosis and honey-combing while ground glass opacities in CTD-ILD is an uncertain pattern and based on available evidence, it was therefore adopted to proceed directly with *Overall Extent of Disease of HRCT* as the single voting item. Note: regarding an end-point *for Overall Extent of Disease on HRCT* in CTD-ILD, no voting option reached the voting threshold of 70%.

Lung Imaging in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
Overall Extent of Lung Disease on HRCT		100% (1/11)	100% (10/10)	100% (1/1)
Possible Primary End-Point		8% (1/12)	9% (1/11)	0% (0/1)
Possible Secondary End-Point		33% (4/12)	27% (3/11)	100% (1/1)
Endpoint Perceived as Difficult to Assign At This Time		58% (7/12)	64% (7/11)	0% (0/1)
End-Point		NONE		

Lung Imaging in CTD ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD ILD Patients**
Overall Extent of Lung Disease on HRCT		92% (23/25)	100% (11/11)	82% (9/11)	91% (20/22)	100% (3/3)
Possible Primary Endpoint		0% (0/23)	0% (0/11)	0% (0/10)	0% (0/21)	0% (0/2)
Possible Secondary Endpoint		65% (15/23)	45% (5/11)	80% (8/10)	62% (13/21)	100% (2/2)
Endpoint Perceived as Difficult to Assign At This Time		35% (8/23)	55% (6/11)	20% (2/10)	38% (8/21)	0% (0/2)
End Point		NONE				

Lung Physiology / Function in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
FVC		100% (11/11)	100% (10/10)	100% (1/1)
FVC as Possible Primary Endpoint		82% (9/11)	80% (8/10)	100% (1/1)
DLCO		100% (11/11)	100% (10/10)	100% (1/1)
DLCO as Possible Secondary Endpoint		91% (10/11)	90% (9/10)	100% (1/1)
Supplemental O2		0% (0/11)	0% (0/10)	0% (0/1)
6MWT Max Desat		45% (5/11)	40% (4/10)	100% (1/1)
6MWT Distance		45% (5/11)	40% (4/10)	100% (1/1)

Lung Physiology / Function in CTD-ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD-ILD Patients**
FVC		100% (24/24)	100% (10/10)	100% (11/11)	100% (21/21)	100% (3/3)
FVCas Possible Primary Endpoint		88% (21/24)	80% (8/10)	100% (11/11)	90% (19/21)	67% (2/3)
DLCO		100% (21/23)	100% (10/10)	80% (8/10)	90% (18/20)	100% (3/3)
DLCO as Possible Secondary Endpoint		87% (20/23)	89% (8/9)	91% (10/11)	90% (18/20)	67% (2/3)
Supplemental O2		4% (1/23)	0% (0/10)	10% (1/10)	5% (1/20)	0% (0/3)
6MWT Max Desat		42% (10/24)	40% (4/10)	36% (4/11)	38% (8/21)	67% (2/3)

Survival: During the NGT, it was proposed by the Survival Team and agreed upon by the assembled group, that *Time to Death* and *Progression Free Survival* in both IPF and CTD-ILD should immediately be tabled to *Research Agenda*, there was no dissension to this. The group opted to proceed, upon advisement of the Survival Team, directly to *All Cause Mortality* and *FVC as a Surrogate Endpoint for Survival* as the voting items.

Survival in IPF

Instrument		**Total for Acceptance**	**Pulms**	**IPF Patient**
All Cause Mortality as a Secondary Endpoint		92% (11/12)	100% (11/11)	0% (0/1)
All Cause Mortality as a Possible Primary Endpoint		25% (3/12)	18% (2/11)	100% (1/1)
FVC as Surrogate Endpoint for Survival		45% (5/11)	40% (4/10)	100% (1/1)
Time to Death	RESEARCH			
Progression Free Survival	RESEARCH			

Survival in CTD ILD

Instrument		**Total for Acceptance**	**Pulms**	**Rheums**	All Physicians	**CTD ILD Patients**
All Cause Mortality as a Secondary Endpoint		92% (23/25)	100% (11/11)	91% (10/11)	95% (21/22)	67% (2/3)
All-Cause Mortality as a Possible Primary Endpoint		0% (0/25) ()1/1	0% (0/11)	0% (0/11)	0% (0/22)	0% (0/3)
FVC as a Surrogate End-Point for Survival		33% (8/24)	30% (3/10)	27% (3/11)	29% (6/21)	67% (2/3)
Progression Free Survival	RESEARCH AGENDA					

Additional questions posed at the NGT Meeting:

Do you think that the CTD-ILD OMERACT group should recommend the collection of bio-samples (according to published guidelines such as the EULAR-EUSTAR biomarker guidelines) in any multicentre RCT in IPF and CTD-ILD? 23/23 Yes by Vote of Hands From All Groups.

The Instruments accepted by the NGT are approved as research agenda items. 23/23 Yes by Vote of Hands From All Groups.

VI. Further References

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