**Background:** UIP is the most common form of chronic interstitial lung disease, comprising up to 70% of cases (1). It can occur idiopathically (I/UIP) or in the setting of connective tissue disease (CTD/UIP). Patients with CTD have a better prognosis than those without, although both populations often undergo lung transplantation (2). Prior studies have demonstrated a preponderance of the NSIP pattern in CTD patients, or even coexisting NSIP and UIP (3-4). We aim to evaluate the utility of finding coexisting multi-lobar NSIP and UIP as a tool to aid in the differentiation between I/UIP and CTD/UIP.

**Design:** With IRB approval, explanted lung tissue from 8 patients (4 I/UIP, 4 CTD/UIP) was identified using an existing database: 2 I/UIP patients with bilateral explants, 1 with right and 1 with left-sided explants; 4 CTD-UIP patients with bilateral explants. Slides from all lobes were evaluated for the presence of NSIP in areas away from fibrosis/honeycombing.

**Result:** 1 of 4 I/UIP patients had cellular NSIP in all lobes and 1 had patchy fibrotic NSIP. 3 of 4 CTD/UIP patients had cellular NSIP in all lobes. In both groups, patients without multi-lobar NSIP demonstrated widened alveolar walls only in the areas adjacent to fibrosis, which was not seen in areas distant from fibrosis.

**Conclusion:** Although the presence of coexisting multi-lobar cellular NSIP is not exclusive to CTD/UIP patients, it does occur with a higher prevalence (25% in I/UIP versus 75% in CTD/UIP), and therefore may serve as a clue to the presence of CTD. Common to both groups are widened alveolar walls near areas of fibrosis, which may mimic fibrotic NSIP. Hence, alveoli near fibrotic/honeycombed areas cannot be used to evaluate for NSIP, and pathologists must be strict in examining pulmonary parenchyma at some distance from typical UIP fibrosis.

**References:**