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| **SUPPLEMENT 1.** Histological changes of 16 cases with SSC-CIP |
| **No.** | **Sex** | **Age** | **Liver** **Histology**  |  | Timefirst signs of chole-stasistohistology(days) |  |
|  | Portal tracts | Bile ducts | Medium/or large hilarbile ducts |
| **biopsy** | **explant** | Fibrosis/ Cirrhosis | Edema/Inflammation | Destructionof bile ducts | Proliferation (ductular reaction) | Periductal fibrosis | Canalicular cholestasis |  |
| 1 | F | 63 | - | + | Liver explant | 189 | ppF | +/Inflammation |  | + | + | + | Medium-sized bile ducts with focal necrosis of epithelium and periductal fibrosisLarge hilar bile ducts with complete necrosis of epithelium |
| 2 | M | 25 | + | - | Liver biopsy | 351 | ppF | +/Edema+/Inflammation | + | + |  |  |  |
| 3 | M | 43 | - | - | - |  | - |  |  |  |  |  |  |
| 4 | M | 26 | - | - | - |  | - |  |  |  |  |  |  |
| 5 | M | 49 | + | + | Liver biopsy+Liver explant | 86 | BiopsyppF |  |  |  |  |  |  |
| 961 | Liver explant:LC | +/Inflammation | +Mild alteration of bile duct epithelium | +Numerousbile ductular proliferations | + | + |
| 6 | M | 33 | - | + | Liver explant | 1718 | LC | +/Inflammation |  | + | + |  |  |
| 7 | M | 53 | - | + | Liver explant | 1402 | LC |  |  | + |  |  |  |
| 8 | M | 57 | - | - |  |  | - |  |  |  |  |  |  |
| 9 | M | 18 | - | - | e.l. |  | - |  |  |  |  |  |  |
| 10 | F | 46 | - | + | Liver explant | 1465 | LC | +/Inflammation | +Vanishingbile duct syndrome |  |  |  |  |
| 11 | F | 60 | + | + | Liver biopsy+Liverexplant | 126 | Biopsy:Portal fibrosis | +/Inflammation |  | +Bile ductular proliferation |  |  |  |
| 237 | Liver explant:ppF | +/Inflammation predominantly lymphocytes | +Ductopenia | +Ductular reaction | Large hilar bile ducts with focal necrosis of epithelium+ Infiltration of epithelium with leucocytes+periductal fibrosis |
| 12 | M | 63 | + | + | Liver biopsy+Liver explant | 29 | Biopsy:Portal fibrosis  | +/Inflammation |  | +Numerousbile ductular proliferations |  | + |  |
| 967 | Liver explant:LC |  | +Numerousbile ductular proliferations | + |
| 13 | F | 47 | + | + | Liver biopsy+Liver explant | 118 |  | +/Inflammation |  |  |  |  |  |
| 1269 | Liver explant:LC |  |
| 14 | M | 57 | - | - |  |  | - |  |  |  |  |  |  |
| 15 | M | 34 | + | + | Liver biopsy+Liver explant | 22 |  | +/Edema+/Inflammation | +Necrosis of bile duct epithelium |  |  | + |  |
| 1001 | Biopsy: Portal fibrosis |
| 1476 | LC | +/Inflammation | +Portal tracts with total loss of bile ducts | +Bile ductular proliferations |
| 16 | F | 60 | - | - | Autopsy |  | LC |  |  | +Bile ductular proliferations |  |  |  |
| Age in years, M= male, F= female, ppF=porto-portalis bridging fibrosis, LC= Liver cirrhosis |