

# SECONDARY SCLEROSING CHOLANGITIS IN CRITICALLY ILL PATIENTS (SSC-CIP) ON EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO)

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# LEARNING OBJECTIVES

The aims of our educational poster are to:

- Define SSC-CIP and provide a brief overview of extracorporeal membrane oxygenation (ECMO) therapy
- Describe the pathogenesis of SSC-CIP and its clinical course
- Illustrate the spectrum of imaging findings found in SSC-CIP by using case examples
- Discuss the management of SSC-CIP
- Highlight the key features of SSC-CIP that radiologists should be aware of.

BACKGROUND

# INTRODUCTION: WHAT IS SSC-CIP?

- Secondary sclerosing cholangiopathy (SSC) comprises a group of inflammatory cholestatic disorders affecting the intra- and extrahepatic biliary ducts leading to stricture formation, biliary cirrhosis and subsequent liver failure.
- Secondary sclerosing cholangitis in critically ill patients (SSC-CIP) was first described in 1997<sup>1</sup> and is now becoming an increasingly recognized form of sclerosing cholangiopathy.<sup>2</sup>
- SSC-CIP should be considered in ITU patients with newly deranged liver function tests who have no prior history of liver disease and no other cause of biliary obstruction.<sup>2</sup>

1) Schmitt M, Kolbel CB, Muller MK, Verbeke CS, Singer MV. Sclerosing cholangitis after burn injury. Z Gastroenterol. 1997;35(10):929-934.

2) Gudanason H, Bjornsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.

# INTRODUCTION: PREVALENCE OF SSC-CIP

- Exact prevalence is difficult to determine as the disease remains under recognised and under reported.
- Leonhardt et al reported 0.61% (16/2633) of all liver transplantations in their centre were due to SSC-CIP.<sup>1</sup>
- Some series have reported a male preponderance (mean M:F ratio = 4.5:1) whilst others have found no significant difference between gender.<sup>1,2</sup>

1) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.

2) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. *Viszeralmedizin* 2015;31:178-184.

# INTRODUCTION: TREATMENT & PROGNOSIS

- SSC-CIP leads to rapid progression of liver cirrhosis; associated with poor survival with up to 50% mortality.<sup>1</sup>
- Liver transplant is the mainstay curative treatment. <sup>1,2</sup>
- Prognosis is poor.
- Transplant-free survival is around 17–40 months, which is lower than in other forms of SSC patients. <sup>2</sup>

1) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.

2) Gudnason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. *Clin Exp Gastroenterol*. 2017; 10: 105–111.

# INTRODUCTION: WHAT IS ECMO?

- Extracorporeal membrane oxygenation (ECMO) is a form of haemodynamic support which facilitates gas exchange.<sup>1,2</sup>
- First used in 1972.<sup>1</sup>
- Increasingly used to manage ITU patients with severe cardiac or respiratory compromise who fail to respond to conventional therapy.<sup>1</sup>
- Two main types of ECMO<sup>1-2</sup>:
  - Veno-arterial (VA) ECMO
  - Veno-venous (VV) ECMO

1) Hosmane SR, Barrow T, Ashworth A, Smith E. Extracorporeal membrane oxygenation: a radiologists' guide to who, what and where. Clin Radiol 2015; 70 (5): e58-66.

2) Lee S, Chatruvedi A. Imaging adults on extracorporeal membrane oxygenation (ECMO). Insights Imaging. 2014; 5 (6):731-742.

# TYPES OF ECMO

## VENO-ARTERIAL ECMO

- Deoxygenated blood drained from vein and oxygenated blood returned into an arterial vessel.
- Complete respiratory & cardiac support
- Peripheral or central VA ECMO
- Indications: cardiogenic shock, post cardio-pulmonary bypass<sup>1,2</sup>
- In central VA ECMO, cannula placement is mediastinal (usually post bypass)<sup>1,2</sup>

## VENO-VENOUS ECMO

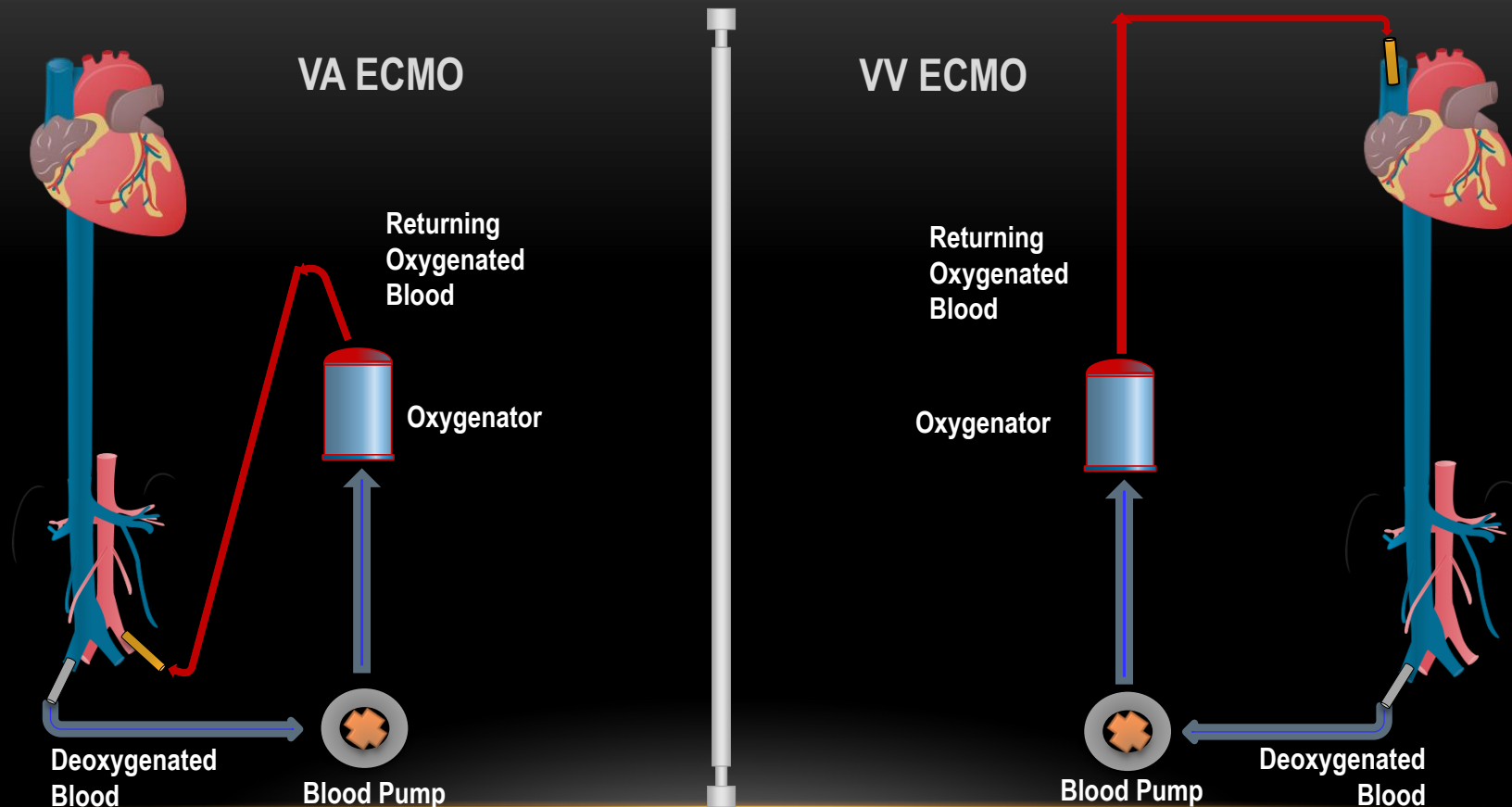
- Deoxygenated blood drained from peripheral vein and oxygenated blood returned into a systemic vein or right atrium<sup>1</sup>.
- Provides respiratory support in patients with adequate cardiac function.
- Indications: Severe pneumonia/sepsis, acute respiratory distress syndrome (ARDS).<sup>1,2</sup>

1) Hosmane SR, Barrow T, Ashworth A, Smith E. Extracorporeal membrane oxygenation: a radiologists' guide to who, what and where. Clin Radiol 2015; 70 (5): e58-66.

2) Lee S, Chatruvedi A. Imaging adults on extracorporeal membrane oxygenation (ECMO). Insights Imaging. 2014; 5 (6):731-742.



Diagram illustrating ECMO circuit in peripheral VA and VV ECMO. ECMO cannulas may have variable positioning.<sup>1-3</sup>



- 1) Hosmane SR, Barrow T, Ashworth A, Smith E. Extracorporeal membrane oxygenation: a radiologists' guide to who, what and where. Clin Radiol 2015; 70 (5): e58-66.
- 2) Lee S, Chatruvedi A. Imaging adults on extracorporeal membrane oxygenation (ECMO). Insights Imaging. 2014; 5 (6):731-742.
- 3) Images drawn using Edraw Max (Version 9.1) [Software] . Available at: <https://www.edrawsoft.com/vector-human-organs.php>

# ECMO FOR RADIOLOGISTS

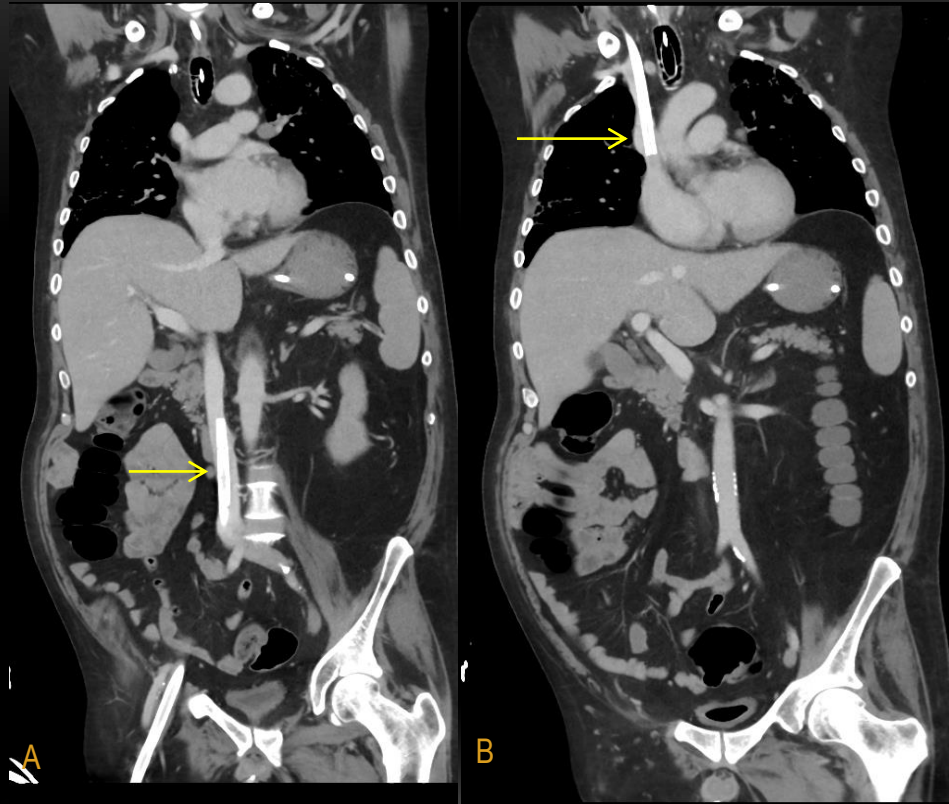
Table illustrating ideal cannula/catheter positions in each type of ECMO<sup>1-2</sup>.

ECMO type	Venous Drainage catheter	Return catheter
VA Central	Right atrium	Ascending aorta with tip pointing upwards
VA Peripheral	Distal IVC/ SVC before cavoatrial junction	Proximal femoral, axillary or subclavian artery
VV Dual Cannula	Distal IVC below hepatic veins via femoral vein	Right atrium via same or opposite femoral vein
VV Dual Cannula	Distal IVC below hepatic veins via femoral vein	Distal SVC/right atrium via internal jugular vein
VV Dual lumen single cannula	Tip advanced to IVC via internal jugular vein Simultaneous drainage of blood from SVC and IVC; blood returned into right atrium	

1) Hosmane SR, Barrow T, Ashworth A, Smith E. Extracorporeal membrane oxygenation: a radiologists' guide to who, what and where. Clin Radiol 2015; 70 (5): e58-66.

2) Lee S, Chatruvedi A. Imaging adults on extracorporeal membrane oxygenation (ECMO). Insights Imaging. 2014; 5 (6):731-742.

# VENO-VENOUS (VV) ECMO



Coronal CT images depicting VV ECMO lines. Right femoral drainage cannula (A) tip is situated in the IVC below hepatic veins and the internal jugular line return cannula (B) is in the SVC.

# INTRODUCTION: PATHOGENESIS OF SSC-CIP

- Progressive and irreversible biliary duct destruction with cholangiocyte damage initiated within few days of ITU admission.<sup>1-4</sup>
- Two different pathophysiological processes have been postulated:
  - an “ischaemic cholangiopathy” hypothesis causing biliary epithelium necrosis vs.
  - “toxic bile” damage theory.<sup>1-4</sup>

1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.

2) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199-4205.

3) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. Ann Hepatol. 2015 Sep-Oct;14(5):695-701.

4) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

# PATHOGENESIS OF SSC-CIP

## ISCHAEMIC THEORY – favoured theory

Cholangiocytes are susceptible to ischaemia due to the nature of their vascular supply (see next slide).<sup>1-3</sup>

ITU patients often exposed to prolonged episodes of hypotension and unique to our department placed on ECMO therapy. These interventions impair flow to the hepatic artery and peribiliary plexus leading to cholangiocyte necrosis.

- 1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.
- 2) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199–4205.
- 3) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

## ISCHAEMIC THEORY – favoured theory

Predilection for intrahepatic biliary duct involvement (Leonhardt et al<sup>1</sup> described 81% patients had only intrahepatic ducts affected compared to 19% which had both intra-and extrahepatic ductal involvement).

This is likely due to sole blood supply from the hepatic artery (see table below)<sup>2-4</sup>

Cell type	Vascular Supply
Intrahepatic duct	Network of small vessels branching from the right & left hepatic arteries. Form a plexus of arterioles, venules, and capillaries within the peribiliary adventitia & peribiliary capillary plexus within the wall
Extrahepatic duct	feeding branches from an average eight arteries (gastroduodenal, right & left hepatic arteries etc)
Hepatocyte	dual supply from portal vein and hepatic artery

- 1) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. Medicine 2015;94(49):e2188.
- 2) Gudanason H, Bjornsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.
- 3) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199-4205.
- 4) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

# PATHOGENESIS OF SSC-CIP

## 'TOXIC BILE' theory

Cholangiocyte damage caused by alteration in balance between bile acids and protective mechanisms. Sepsis and severe hypotension believed to alter this balance.<sup>1-3</sup>

Detergent properties of bile acids destroy the lipid cellular membrane of cholangiocytes leading to sclerosing cholangitis.

Postulated that individuals with genetic variants of transporter lipid export pump MDR3 have higher risk of toxic bile formation when exposed to hypoxia or inflammatory stress. Thought to explain why some ITU patients develop SSC-CIP whilst others recover without biliary deterioration.<sup>1</sup>

- 1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.
- 2) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199-4205.
- 3) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

# RISK FACTORS FOR SSC-CIP

- Severe haemodynamic instability, mechanical ventilation, increased intraperitoneal fat volume, prone positioning and hypercoagulable state associated with increased risk of developing SSC-CIP <sup>1-4</sup>
- Polytrauma patients and patients who underwent major surgery reported to be at higher risk. <sup>3</sup>
- Limited data in the literature on SSC-CIP in ECMO patients
- Akbar et al <sup>5</sup> reported a case of ischaemic cholangiopathy following ECMO therapy
- Small study by Weig et al <sup>4</sup> showed 60% of SSC-CIP patients received ECMO therapy versus 44% of non-SSC patients; although not statistically significant

1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.

2) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199-4205.

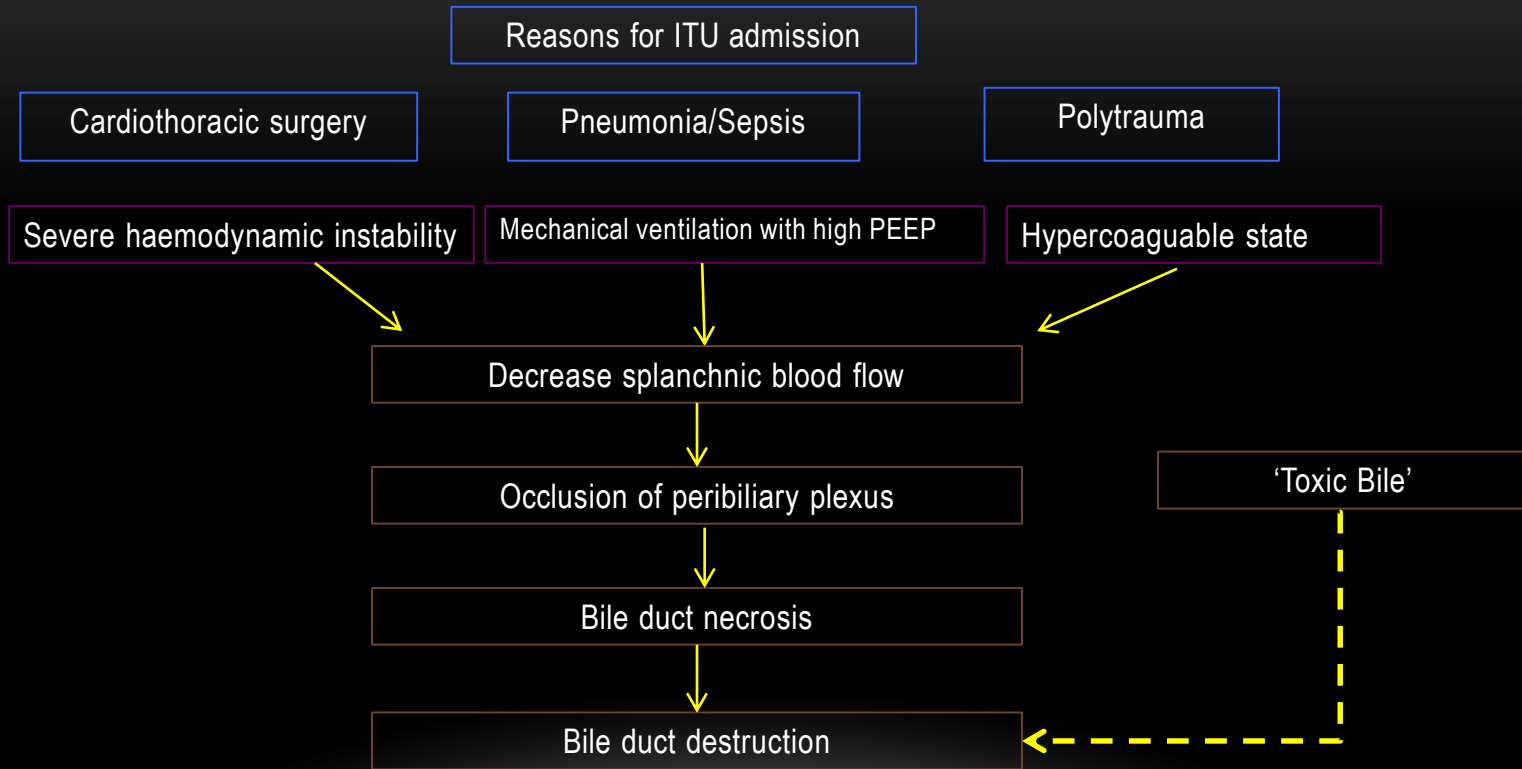
3) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

4) Weig et al. Abdominal Obesity and prolonged prone positioning increase risk of developing sclerosing cholangitis in critically ill patients with influenza A-associated ARDS. European Journal of Medical Research 2012 17:30.

5) Akbar A, Baron TH. Ischemic biliary injury following extracorporeal membrane oxygenation (ECMO). Dig Liver Dis 2012; 44 (8):705.



# PATHOGENESIS SUMMARY<sup>1-4</sup>



- 1) Gudnason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.
- 2) Benninger J, Grobholz R, Oeztuerk Y, Antoni CH, Hahn EG, Singer MV. Sclerosing cholangitis following severe trauma: description of a remarkable disease entity with emphasis on possible pathophysiologic mechanisms. World J Gastroenterol 2005; 11:4199-4205.
- 3) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.
- 4) Voigtländer T, Jaeckel E, Lehner F, Manns M, Lankisch T. Liver transplantation for critically ill patients with secondary sclerosing cholangitis: Outcome and complications. Liver Transplantation 2015; 21:1295–1299.

# CLINICAL PRESENTATION

- Acute derangement of liver function with a cholestatic picture is the first indicator.<sup>1-5</sup>
- Comparatively serum alanine aminotransferase (ALT) and aspartate aminotransferase demonstrate moderate elevation.<sup>2</sup>
- Clinical symptoms are rare; although Leonhardt et al<sup>2</sup> reported profound weight loss (mean 18 kg) in 94% of patients.

1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105-111.

2) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. Medicine 2015;94(49):e2188.

3) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. Viszeralmedizin 2015;31:178-184.

4) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. Ann Hepatol. 2015 Sep-Oct;14(5):695-701.

5) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

# CLINICAL PRESENTATION

Table illustrating pattern of liver enzyme derangement<sup>1-5</sup>.

Liver enzyme	Peak (Mean time)	Level (x normal)
Gamma-glutamyl transferase (GGT)	First to elevate, 7 days after event	20 to 50 times
Alkaline phosphatase (ALP)	Within few days of GGT	5 to 21 times
Bilirubin	16 days	3 to 39 times

- 1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105-111.
- 2) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. Medicine 2015;94(49):e2188.
- 3) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. Viszeralmedizin 2015;31:178-184.
- 4) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. Ann Hepatol. 2015 Sep-Oct;14(5):695-701.
- 5) Leonhardt et al. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. Crit Care. 2015 Mar 31;19:131.

# IMAGING FEATURES/DETAILS

# WHAT IS THE ROLE OF RADIOLOGY?

- Ultrasound: diagnostic accuracy approximately 30% compared to ERCP<sup>1</sup>
- ITU patients (especially if on ECMO therapy) often undergo CT assessment which can detect early changes such as intrahepatic biliary dilatation
- ERCP is considered by some series as 'gold standard' for diagnosis of SSC-CIP. <sup>1-3</sup> However, MRCP provides a non-invasive overview of the biliary system and can visualise parts of the biliary tree not opacified during ERCP due to strictures.<sup>4</sup>
- Liver biopsy: variable histology depending on stage of SSC-CIP. Studies on explanted livers describe features of an ischaemic cholangiopathy. <sup>2,5</sup> Limited diagnostic value with no definite correlation between histological features, liver function tests and outcome<sup>1</sup>.

1) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. *Viszeralmedizin* 2015;31:178-184.

2) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.

3) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. *Clin Exp Gastroenterol*. 2017; 10: 105–111.

4) Vitellas et al. Radiologic manifestations of sclerosing cholangitis with emphasis on MR cholangiopancreatography. *RadioGraphics* 2000; 20:959-975.

5) Voigtländer T, Jaeckel E, Lehner F, Manns M, Lankisch T. Liver transplantation for critically ill patients with secondary sclerosing cholangitis: Outcome and complications. *Liver Transplantation* 2015; 21:1295–1299.

# STAGES OF SSC CIP

Three stages of SSC-CIP described based on endoscopic retrograde cholangiopancreatography (ERCP) findings. <sup>1-4</sup>

- Stage 1/Early: Extensive 'ribbon-like' intraductal filling defects known as 'biliary casts' form in the biliary tree. Biliary casts reported in 87%-93% of patients and considered pathognomonic for SSC-CIP<sup>2,3</sup>
- Stage 2/Intermediate: Rapid progressive intrahepatic bile duct destruction manifesting as multiple irregular strictures, beading and dilatations.
- Stage 3/Late: Due to progressive destruction, intrahepatic bile duct branches are obliterated, leaving behind rudimentary central biliary system giving the picture of a 'pruned tree'.

1) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. Clin Exp Gastroenterol. 2017; 10: 105–111.

2) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. Medicine 2015;94(49):e2188.

3) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. Viszeralmedizin 2015;31:178-184.

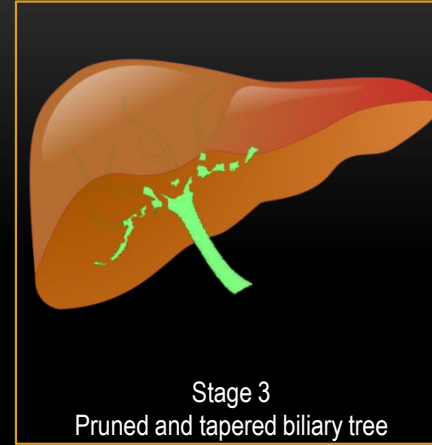
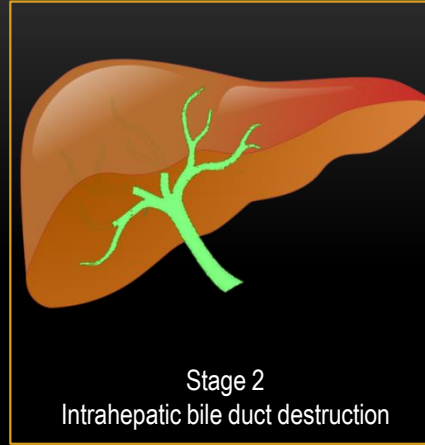
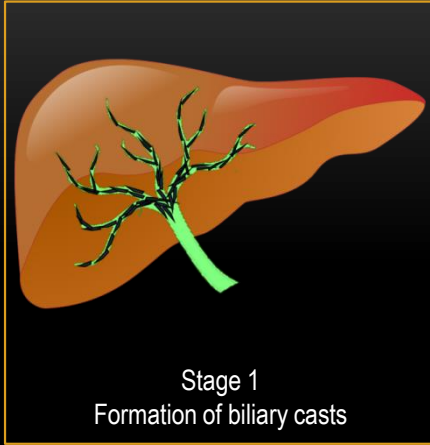
4) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. Ann Hepatol. 2015 Sep-Oct;14(5):695-701.

Stages of SSC-CIP: Video below illustrates the 3 progressive stages.<sup>1,2</sup>



- 1) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.
- 2) Images drawn using Edraw Max (Version 9.1) [Software] . Available at: <https://www.edrawsoft.com/vector-human-organs.php>

## Stages of SSC- CIP<sup>1,2</sup>



ERCP image depicting  
Stage II SSC-CIP -  
destruction of the  
intrahepatic biliary tree.

- 1) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.
- 2) Images drawn using Edraw Max (Version 9.1) [Software] . Available at: <https://www.edrawsoft.com/vector-human-organs.php>



# IMAGING FEATURES

Imaging manifestations include:

- Ultrasound (US): intrahepatic +/- extrahepatic biliary dilatation; US features of liver cirrhosis may be seen within 6 months of the onset of SSC-CIP. <sup>1</sup>
- CT: biliary dilatation with adjacent formation of 'bile lakes' due to biliary obstruction from cast formation. Some may develop multiple biliary microabscesses which contact the biliary tree. <sup>2</sup>
- MRCP: diffuse intrahepatic biliary duct irregularity, wall, thickening, multifocal strictures and dilatations. <sup>3,4</sup>
- ERCP: Biliary casts seen as intraductal filling defects; similar findings to MRCP with pruned appearance in later stages. <sup>2</sup>

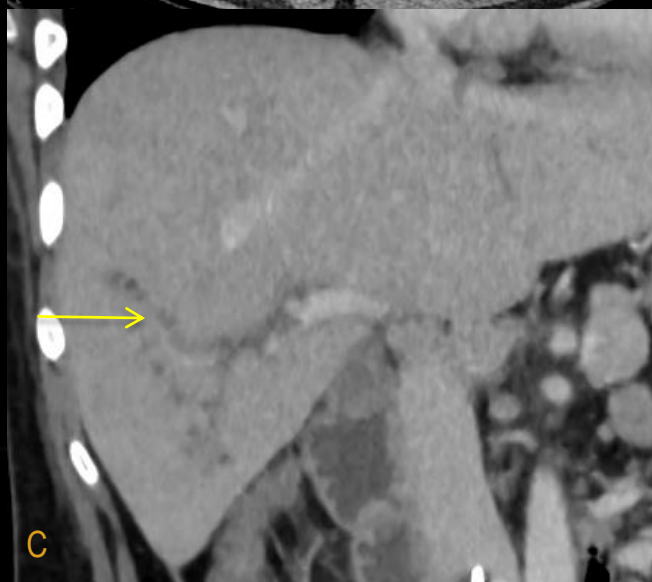
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2) Gudanason H, Bjornsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. *Clin Exp Gastroenterol*. 2017; 10: 105–111.

3) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. *Viszeralmedizin* 2015;31:178-184.

4) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. *Ann Hepatol*. 2015 Sep-Oct;14(5):695-701.

## CASE 1



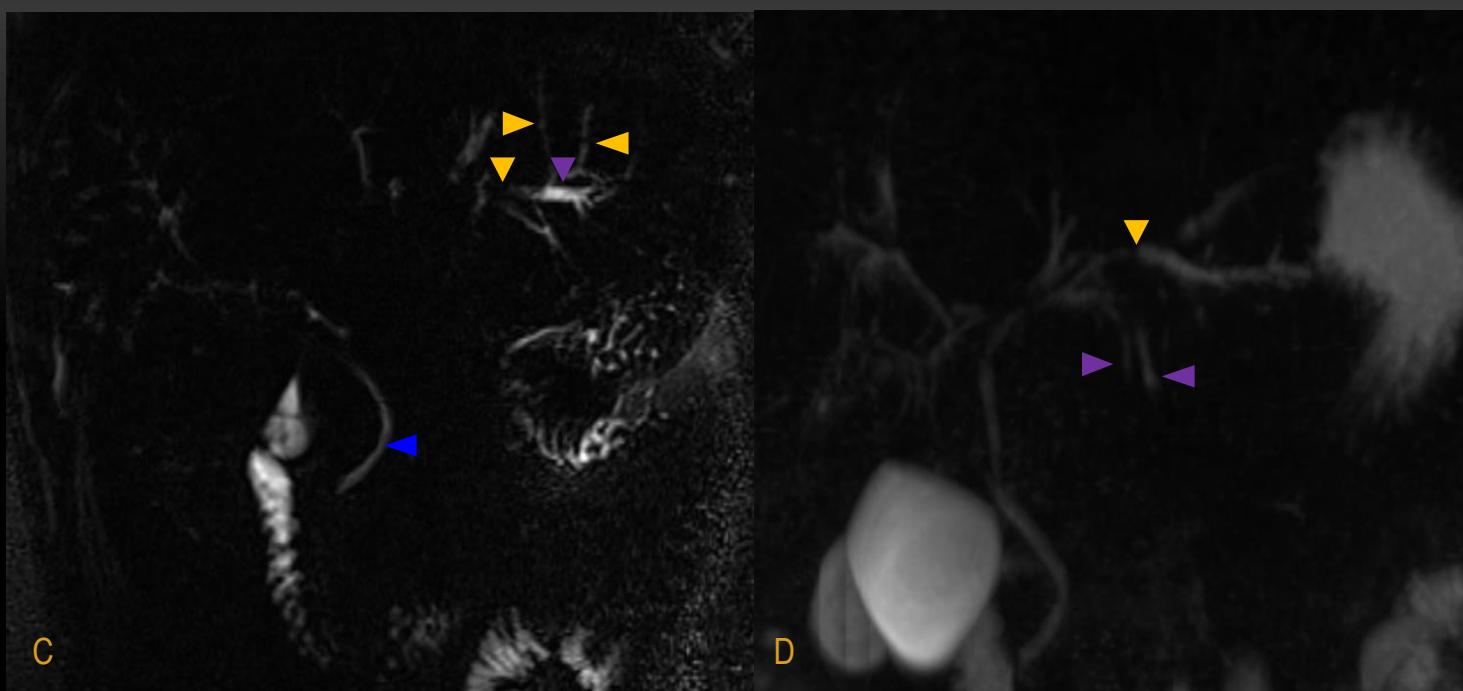
Adolescent male previously fit and healthy polytrauma patient, commenced on ECMO.

(A) Initial trauma CT: extensive splenic injury. Note normal liver.

(B & C) CT post splenectomy and 2 months post-initial CT. Bloods showed Bilirubin 8 (<20 IU/L), raised ALP 1530 (30-130 IU/L) and ALT 87 (<55 IU/L). No GGT.

Coronal CT image (B) demonstrates new intrahepatic biliary dilatation and terminal ductal hypodensities. Axial CT (C) confirms these focal hypodensities as 'bile lakes' in communication with the intrahepatic biliary tree.

## CASE 1



(C) MRCP at similar time to CT demonstrates multiple irregular strictures (yellow arrowhead) leading to segmental intrahepatic biliary dilatation (purple arrowhead), and normal CBD (blue arrowhead).

(D) Follow up MRCP 18 months later demonstrates persistent stricturing (yellow arrowhead) and upstream dilatation of the segment 2 ducts. Also note the pruned appearance of the intrahepatic biliary tree (purple arrowheads). The previously seen bile lakes seen on CT have resolved.

Patient survived admission and was discharged under care of hepatologist.

## CASE 2



Male patient presented with sepsis secondary to severe pneumonia and commenced on VV ECMO.

(A) Normal liver on initial CT.

(B) & (C) Portal venous phase axial and coronal CT images at time of newly deranged LFTs\* demonstrate dilated intra-hepatic biliary tree and small hypo-densities suggestive of bile 'lakes'.

\*raised bilirubin 371 (<20 IU/L), ALP 456 (30-130 IU/L) and ALT 80 (<55 IU/L). GGT not routinely performed at our institution.

## CASE 2

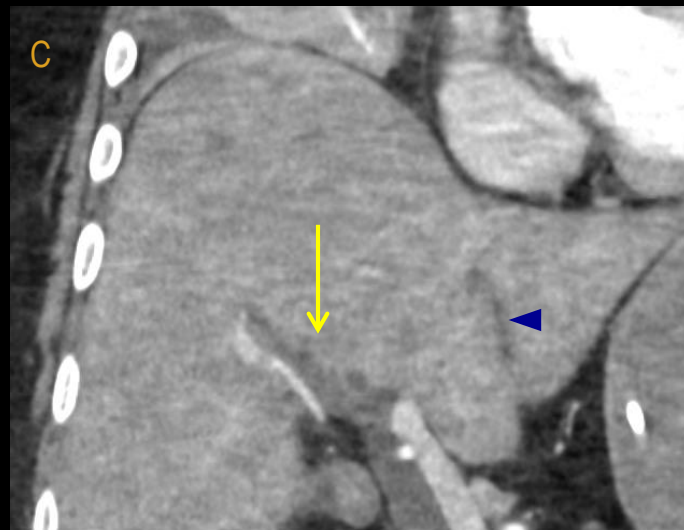
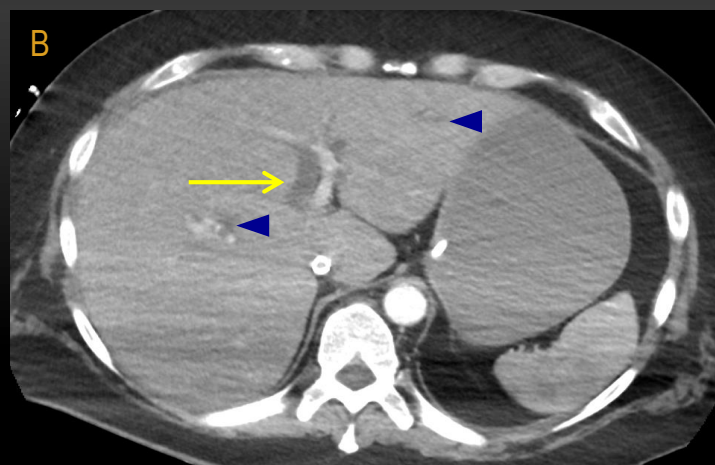
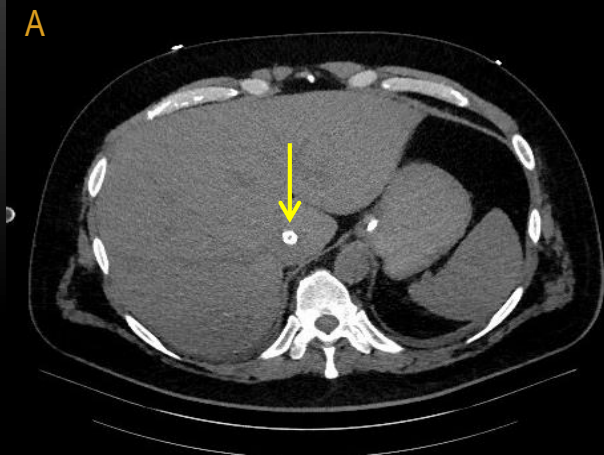


Subsequent ERCP demonstrated extensive destruction of the intrahepatic biliary tree.

Patient unfortunately died during ITU admission.



## CASE 3



Male patient with severe sepsis (klebsiella and flu) commenced on VV ECMO.

(A) Initial axial CT showed normal liver and VV ECMO catheter in the intrahepatic IVC (yellow arrow)

(B & C) Further CT following deterioration of LFTs\*  
CT showed increasing CBD dilatation and new intrahepatic biliary dilatation (blue arrowheads)

Patient unfortunately died during ITU admission.

\*(Bilirubin 43, ALP 1127, ALT 67).

# COMPLICATIONS OF SSC-CIP

- As the necrotic cells slough from the biliary ductal walls, adherent biliary casts form which obstruct the intrahepatic ducts<sup>1-3</sup> (see diagram on next slide)
- Increased risk of developing bacterial cholangitis/cholangiosepsis. <sup>1-3</sup> Enterococcus faecium reported as one of the associated pathogens.<sup>4</sup>
- Some series found acalculous cholecystitis to occur in 50% of patients with SSC-CIP and of these half required emergency cholecystectomy. <sup>2,3</sup>

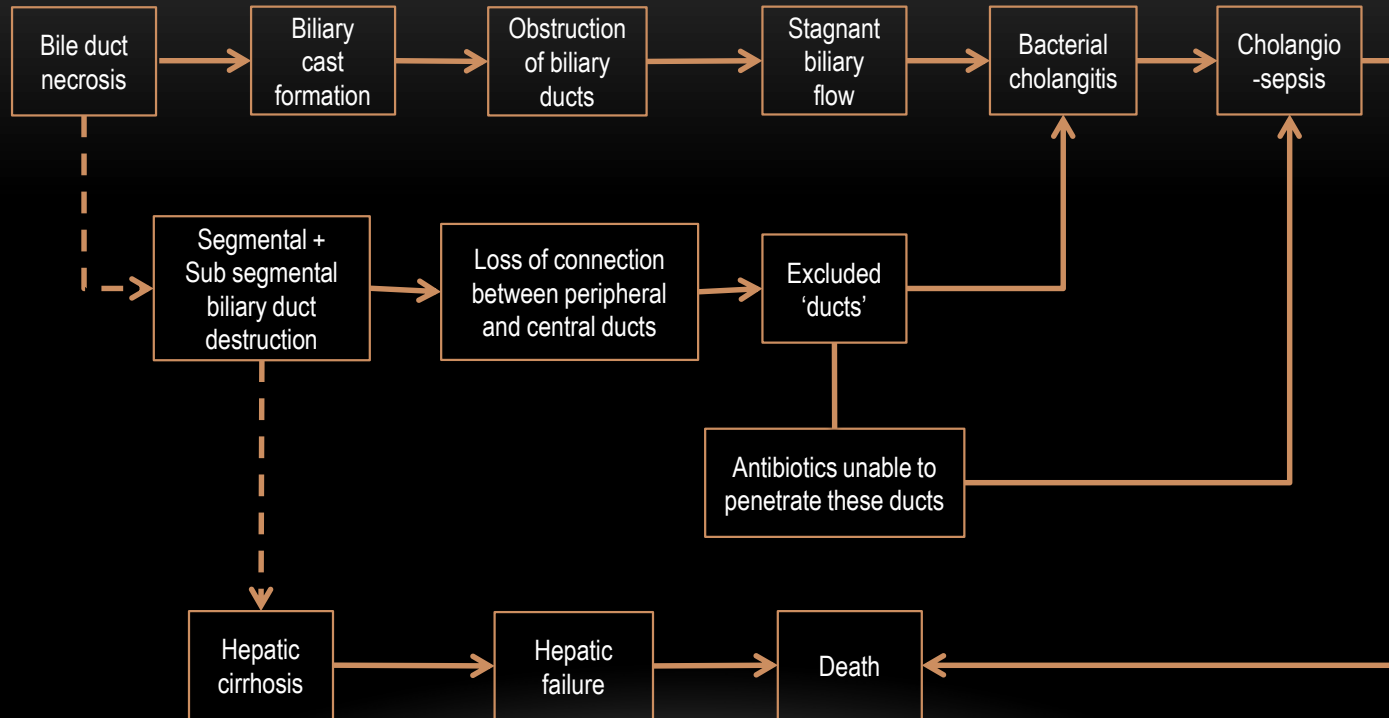
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3) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. Medicine 2015;94(49):e2188.

4) Gelbmann CM, Rümmele P, Wimmer M, Hofstaedter F, Göhlmann B, Endlicher E Ischemic-like cholangiopathy with secondary sclerosing cholangitis in critically ill patients. Am J Gastroenterol 2007; 102:

# Diagram illustrating the complications in SSC-CIP



- 1) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. *Viszeralmedizin* 2015;31:178-184.
- 2) Gudanason H, Björnsson ES. Secondary sclerosing cholangitis in critically ill patients: current perspectives. *Clin Exp Gastroenterol*. 2017; 10: 105–111.
- 3) Leonhardt et al. Secondary Sclerosing Cholangitis in Critically Ill Patients: Clinical Presentation, Cholangiographic Features, Natural History, and Outcome: A Series of 16 Cases. *Medicine* 2015;94(49):e2188.
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# MANAGEMENT

- Limited treatment options but various studies have explored endoscopic intervention:<sup>1-3</sup>
  - Endoscopic sphincterotomy, dilatation of strictures, intermittent stenting (risk of stent occlusion & cholangitis)
  - Removal of biliary casts in larger intrahepatic biliary ducts with dormia basket<sup>1</sup>
  - Sludge extraction
  - Continuous saline rinsing through a nasobiliary drainage for several days for smaller bile ducts (lower success rate due to adherent/insoluble biliary casts)<sup>1-2</sup>
- Some report short term biochemical improvement when endoscopic treatment is combined with ursodeoxycholic acid (UDCA).<sup>2</sup>
- However, progressive biliary duct damage cannot be prevented.
- Liver transplantation is the only curative treatment.<sup>4</sup>

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3) Ben-Ari Z et al. Secondary sclerosing cholangitis following major burn. Ann Hepatol. 2015 Sep-Oct;14(5):695-701.

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# PROGNOSIS

- Poor clinical outcome either due to complications from SSC-CIP such as sepsis or liver cirrhosis<sup>1,2</sup>
- Irreversible with high mortality and morbidity
- Median survival is shorter in patients with SSC-CIP (13 months) compared to other forms of secondary sclerosing cholangitis (72 months) and primary sclerosing cholangitis (89 months)<sup>2</sup>
- Studies report 1-year survival without liver transplantation between 42 to 55%<sup>2</sup>
- No reported cases of hepatic/biliary malignancy in SSC-CIP patients in current literature likely due to short life expectancy and lack of long term follow up<sup>1</sup>

1) Kirchner G.I. Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. Viszeralmedizin 2015;31:178-184.

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# KEY LEARNING POINTS:

- With rapid advancements in intensive care, SSC-CIP is emerging as an important differential for acute liver impairment in ITU patients.
- Has an insidious clinical presentation with first biochemical abnormality usually being elevated GGT.
- On imaging, development of new intrahepatic biliary dilatation +/- bile lake formation should raise clinical suspicion of SSC-CIP in patients with previously normal liver.
- Biliary cast formation on ERCP is felt to be pathognomonic of SSC-CIP.
- SSC-CIP has poor prognosis with liver transplantation being the definitive treatment.
- Prompt diagnosis is vital so early contact with transplant centres can be initiated.

# CONCLUSION

- In our poster, we have summarised the clinical and radiological features of SSC-CIP that radiologists should be aware of
- Radiologists play a key role in the early detection of SSC-CIP
- Increased awareness is vital in guiding clinical management
- Little data is available in the literature on SSC-CIP in the ECMO setting
- To our knowledge, we are one of few institutions to present imaging findings of SSC-CIP observed in the ECMO subset.