



Canadian
Biliary Atresia
Registry

Registre
canadien
d'atrésie des voies biliaires

Wishing you Happy Holidays!
Thank you all for your contributions and continued support to CBAR throughout the years. We hope everyone has a safe and amazing holiday season.



News in the CBAR Community **New BC Children's Hospital Co-Site Investigator** **— Dr. Robert Baird**

We would like to welcome Dr. Robert Baird as the new CBAR co-site investigator for BC Children's Hospital (BCCH). He will be working with the principal site investigator at BCCH, Dr. Richard Schreiber, to maintain the CBAR database and support CBAR based initiatives.

Biliary Atresia Workup Algorithm

The CBAR Steering Committee is working on a biliary diagnostic algorithm in an effort to improve standards of care and outcome for Biliary Atresia in Canada. Stay tuned for further information in 2019.

American Association for the Study of Liver Disease (AASLD) 2018 Meeting

Biliary atresia abstracts of interest:

- *Abstract 15:* Predictors of outcome in children with biliary atresia surviving with native liver at two years of age
Venkat, VL et al. (2018)
- *Abstract 30:* A molecular signature predicts 2-year outcome of biliary atresia and identifies a therapeutic target to suppress fibrosis
Luo, Z. et al. (2018).
- *Abstract 137:* Prognostic value of serum bile acids after Kasai portoenterostomy in biliary atresia.
Harpavat, S. et al. (2018)
- *Abstract 186:* Diagnostic Accuracy of Serum Matrix Metalloproteinase-7 for Biliary Atresia
Yang, L. et al. (2018)

All articles can be found at:

<https://aasldpubs.onlinelibrary.wiley.com/doi/epdf/10.1002/hep.30256>

CBAR Registry Updates

- **Total Number of Cases Nationally: 62**
- **Cases by year:**
 - **2014: 18 cases**
 - **2015: 19 cases**
 - **2016: 10 cases**
 - **2017: 15 cases**
- **Number of sites entering data: 11 sites**



Recent Publications By CBAR Members

Province-wide Biliary Atresia Home Screening Program in British Columbia: Evaluation of the First 2 Years.

Woolfson, JP; Schreiber, R; Butler, AE; MacFarlane, J; Kaczorowski, J; Masucci, L; Bryan, S; Collet, JP. *J Pediatr Gastroenterol Nutr.* 2018 June; 66(6), 845-849.

Early Posthepatoportoenterostomy Predictors of Native Liver Survival in Biliary Atresia.

Nightingale, S; Stormon, MO; O'Loughlin, EV; Shun, A; Thomas, G; Benchimol, EI; Day, AS; Adams, S; Shi, E; Ooi, CY; Kamath, BM; Fecteau, A; Langer, JC; Roberts, EA; Ling, SC; Ng, VL. *J. Pediatr Gastroenterol Nutr.* 2017, 64, 203-209.

Knowledge Updates

Prognosis of Biliary Atresia after 2-year survival with native liver: a nationwide cohort analysis

Witt, M; van Wessel, DBE; de Kleine, RHJ; Bruggink, JLM; Hulscher, JBF; Verkade, HJ; NeSBAR. *J Pediatr Gastroenterol Nutr.* 2018 Dec, 67(6), 689-694.

Learning curve of laparoscopic Kasai portoenterostomy for biliary atresia: a report of 100 cases

Y, Ji; Yang, K; Zhang, X; Chen, S, Z, Xu. *BMC Surg.* 2018 Nov 26. doi:10.1186/s12893-018-0443y

Impact of steroid therapy on early growth in infants with Biliary Atresia: the multicenter steroids in Biliary Atresia randomized trial

Alonso, EM; Ye, W; Hawthorne, K; Venkat, V; Loomes, KM; Mack, CL; Hertel, PM; Karpen, SJ; Kerkar, N; Molleston, JP; Murray, KF; Romero, R; Rosenthal, P; Schwarz, KB; Shneider, BL; Suchy, FJ; Turmelle, YP; Wang, KS; Sherker, AH; Sokol, RJ; Bezerra, JA; Magee, JC. *J Pediatr.* 2018 Nov. doi:10.1016/j.jpeds.2018.07.002

Biliary-enteric draining vs primary liver transplant as initial treatment for children with Biliary Atresia.

LeeVan, E; Matsuoka, L; Cao, S; Groshen, S; Alexopoulos, S. *JAMA Surg.* 2018 Sep 12; doi:10.1001/jamasurg.2018.3180

Biliary Atresia: Clinical and research challenges for the twenty-first century

Bezerra, JA; Wells, RG; Mack, CL; Karpen, SJ; Hoofnagle, JH; Doo, E; Sokol, RJ. *Hepatology.* 2018 Sept, 68(3), 1163-1173

Factors associated with timing and adverse outcomes in patients with Biliary Atresia undergoing Kasai Hepatoportoenterostomy

Townsend, MR; Jaber, A; Abi Nader, H; Eid, SM; Schwarz, K. *J Pediatr.* 2018 Aug, 199, 237-242.

Japanese Biliary Atresia Registry

Nio, M. *Pediatr Surg Inter.* 2017 Dec, 32(12), 1319-1325.

Incidence of Biliary Atresia and timing of hepatoportoenterostomy in the United States

Hopkins, PC; Yazigi, N; Nylund CM. *J Pediatr.* 2017 Aug, 187, 253-257.

Questions or Concerns?

If you have any questions or concerns about case entry on RedCap, REB, or other issues, please do not hesitate to contact Crystal Ng (CBAR Research Coordinator) at 604-875-2345 x 4313 or crystal.ng@cw.bc.ca