



Happy Holidays!

#### DECEMBER 2021



We have accomplished a lot this year! Home screening card > 100 patients across 13 sites REDCap upgraded Working with families Website updated & now includes french site Abstract presented @ CAPS Manuscript submission in progress

#### **Co-Chairs**

Rick Schreiber – rschreiber@cw.bc.ca Jean-Martin Laberge - jeanmartinlaberge@hotmail.com

#### **National Coordinator**

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### **Steering Committee Members**

Brian Cameron Carolina Jimenez George Anthopoulos Najma Ahmed Natalie Yanchar Sherif Emil Steve Martin

To a healthy, happy & successful 2022



## **Updates**

## Home Stool Screening Program

<u>BC's Biliary Atresia Home</u> <u>Screening program</u> is leading the way.



To implement this program in Ontario, funding via Ontario's Newborn Screen Let's get this approved has been secured.

#### Parent Committee

We have been working with families & hope to have some patient stories to share on our website to help raise awareness. Should you have any interested patients, please contact the National Coordinator.

We will look at how to better communicate, advertise and help support families of children with biliary atresia through resources.

#### Website

### te cbar.ca

Our website cbar.ca has been upgraded. Most the webpages are now also available in French.

#### Retirements

We would like to thank Dr. Cameron & Dr. Martin for their continued efforts and wish them well in their retirement.

We will be recruiting new Steering Committee members in 2022.

# **Progress on CBAR Data**

#### Patients per site

A lot of work has gone into ensuring patient data is accurate and up to date in REDCap. We currently have 109 patients enrolled.





#### REDCap

REDCap has been upgraded with greater security and new features. If you have questions, contact Elena Guadagno.



Abstract Presented at the 2021 Annual meeting for The Canadian Association of Paediatric Surgeons (CAPS)

Contemporary National Biliary Atresia Investigations and Outcomes: A report from the Canadian Biliary Atresia Registry G Eamer, RA Schreiber, S Emil, E Guadagno, D Briatico, BH Cameron and the Canadian Biliary Atresia Registry

Purpose: Biliary atresia (BA) outcomes are improved for infants having Kasai Portoenterostomy (KP) at younger age. Delays in preoperative investigations for infants with jaundice may contribute to worse outcomes. Our study objective was to assess the effect of investigational studies on duration between presentation and KP.

Methods: We analyzed prospectively collected Canadian Biliary Atresia Registry data, focusing on preoperative investigations, duration between presentation and operation, and native liver survival (NLS).

Results: Sixty-three patients had KP at 13 Canadian Children's Hospitals from 2014-2020, at a median age of 52d. Evaluation by a pediatric surgeon occurred at a median age of 41d (n=59), and KP occurred at a median of 11d post-evaluation (Interquartile range [IQR] 8-17d). Liver biopsy, but not HIDA or cholangiogram, was associated with longer time to KP (16.8d vs. 9.2d, p=0.01). Chi square analysis of infants under/over 45 days of age at KP found no difference in rates of pre-operative cholangiograms (p=0.16) or percutaneous biopsies (p=0.6), but fewer HIDA scans were done in older infants (student's t-test p=0.02). Many tests were non-diagnostic (Table 1). NLS at 6, 12 and 24-months was 87% (n=54), 62% (n=50) and 55% (n=47) respectively. KP under 45d trended towards better NLS at 6-months (95% vs. 82%, p=0.12) but not 12 or 24-months.

Conclusion: Preoperative liver biopsy is associated with delay in Kasai portoenterostomy. Many preoperative tests were non-diagnostic. An expeditious BA diagnostic algorithm, especially for infants over 45 days of age at presentation, may decrease time to KP and improve NLS.

# Working on manuscript submission for 2022.







#### DECEMBER 2021

## **Biliary Atresia Publications by CBAR Members**

Schreiber, R. A. (2020). Newborn Screening for Biliary Atresia. Jama, 323(12), 1137-1138. doi:10.1001/jama.2020.2727 PMID:32207779

Masucci, L., Schreiber, R. A., Kaczorowski, J., Collet, J. P., & Bryan, S. (2019). Universal screening of newborns for biliary atresia: Costeffectiveness of alternative strategies. J Med Screen, 26(3), 113-119. doi:10.1177/0969141319832039 PMID:30871409

Woolfson, J. P., Schreiber, R. A., Butler, A. E., MacFarlane, J., Kaczorowski, J., Masucci, L., . . . Collet, J. P. (2018). Province-wide Biliary Atresia Home Screening Program in British Columbia: Evaluation of First 2 Years. J Pediatr Gastroenterol Nutr, 66(6), 845-849. doi:10.1097/mpg.00000000001950 PMID:29570556

Stagg H, Cameron BH, Ahmed N, Butler A, Jimenez-Rivera C, Yanchar NL, Martin SR, Emil S, Anthopoulos G, Schreiber RA, Laberge JM; Canadian Biliary Atresia Registry. (2017). Variability of diagnostic approach, surgical technique, and medical management for children with biliary atresia in Canada - Is it time for standardization? J Pediatr Surg, 52(5), 802-806. doi:10.1016/j.jpedsurg.2017.01.041 PMID:28189446

Schreiber, R. A., & Butler, A. (2017). Screening for biliary atresia: it's in the cards. Can Fam Physician, 63(6), 424-425. PMID:28615387

Butler, A. E., Schreiber, R. A., Yanchar, N., Emil, S., & Laberge, J. M. (2016). The Canadian Biliary Atresia Registry: Improving the care of Canadian infants with biliary atresia. Paediatr Child Health, 21(3), 131-134. PMID:27398049

Schreiber, R. A., Masucci, L., Kaczorowski, J., Collet, J. P., Lutley, P., Espinosa, V., & Bryan, S. (2014). Home-based screening for biliary atresia using infant stool colour cards: a large-scale prospective cohort study and cost-effectiveness analysis. J Med Screen, 21(3), 126-132. doi:10.1177/0969141314542115 PMID:25009198

Chitsaz, E., Schreiber, R. A., Collet, J. P., & Kaczorowski, J. (2009). Biliary atresia: the timing needs a changin'. Can J Public Health, 100(6), 475-477. doi:10.1007/bf03404348 PMID:20209745

Schreiber, R. A., Barker, C. C., Roberts, E. A., Martin, S. R., Alvarez, F., Smith, L., . . . Critch, J. (2007). Biliary atresia: the Canadian experience. J Pediatr, 151(6), 659-665, 665.e651. doi:10.1016/j.jpeds.2007.05.051 PMID:18035148

Schreiber, R. A., & Kleinman, R. E. (2002). Biliary atresia. J Pediatr Gastroenterol Nutr, 35 Suppl 1, S11-16. doi:10.1097/00005176-200207001-00005 PMID:12151815

# Biliary Atresia Screening Publications

Wications Xiao, Y., Zhou, Y., Zhou, K., & Cai, W. (2021). Targeted Metabolomics Reveals Birth Screening Biomarkers for Biliary Atresia in Dried Blood Spots. J Proteome Res. PMID:34850627

> Zhao, D., Gu, S., Gong, X., Li, Y., Sun, X., Chen, Y., . . . Zhang, Y. (2021). Webbased calculator for biliary atresia screening in neonates and infants with cholestasis. Transl Pediatr, 10(2), 225-235. PMID:33708508

Rabbani, T., Guthery, S. L., Himes, R., Shneider, B. L., & Harpavat, S. (2021). Newborn Screening for Biliary Atresia: a Review of Current Methods. Curr Gastroenterol Rep, 23(12), 28. PMID:34817690

Madadi-Sanjani, O., Kuebler, J. F., Uecker, M., Pfister, E. D., Baumann, U., Kunze-Hullmann, B., . . . Petersen, C. (2021). Province-Wide Stool Color Card Screening for Biliary Atresia in Lower-Saxony: Experiences with Passive Distribution Strategies and Results. Int J Neonatal Screen, 7(4). PMID:34842600

Goodhue, C., Fenlon, M., & Wang, K. S. (2017). Newborn screening for biliary atresia in the United States. Pediatr Surg Int, 33(12), 1315-1318. PMID:28983658

Matsui, A. (2017). Screening for biliary atresia. Pediatr Surg Int, 33(12), 1305-1313. PMID:28983697

Harpavat, S., Garcia-Prats, J. A., & Shneider, B. L. (2016). Newborn Bilirubin Screening for Biliary Atresia. N Engl J Med, 375(6), 605-606. PMID:27509119

Harpavat, S., Ramraj, R., Finegold, M. J., Brandt, M. L., Hertel, P. M., Fallon, S. C., . . . Shneider, B. L. (2016). Newborn Direct or Conjugated Bilirubin Measurements As a Potential Screen for Biliary Atresia. J Pediatr Gastroenterol Nutr, 62(6), 799-803. d PMID:26720765

Sokol, R. J., Shepherd, R. W., Superina, R., Bezerra, J. A., Robuck, P., & Hoofnagle, J. H. (2007). Screening and outcomes in biliary atresia: summary of a National Institutes of Health workshop. Hepatology, 46(2), 566-581. PMID:17661405

## Biliary Atresia Reviews & more

Hinojosa-Gonzalez, D. E., Bueno, L. C., Roblesgil-Medrano, A., Salgado-Garza, G., Hurtado-Arellano, S., Farias, J. S., . . . Flores-Villalba, E. (2021). Laparoscopic vs open portoenterostomy in biliary atresia: a systematic review and meta-analysis. Pediatr Surg Int, 37(11), 1477-1487. PMID:34269866

He, L., Ip, D. K. M., Tam, G., Lui, V. C. H., Tam, P. K. H., & Chung, P. H. Y. (2021). Biomarkers for the diagnosis and post-Kasai portoenterostomy prognosis of biliary atresia: a systematic review and meta-analysis. Sci Rep, 11(1), 11692. PMID:34083585

Alexander, E. C., Greaves, W., Vaidya, H. J., Burford, C., Jain, V., & Samyn, M. (2021). Social and Educational Outcomes in Patients with Biliary Atresia: A Systematic Review. J Pediatr Gastroenterol Nutr. PMID:34560722

Bezerra, J. A., Wells, R. G., Mack, C. L., Karpen, S. J., Hoofnagle, J. H., Doo, E., & Sokol, R. J. (2018). Biliary Atresia: Clinical and Research Challenges for the Twenty-First Century. Hepatology, 68(3), 1163-1173. PMID:29604222

Bezerra, J. A., Wells, R. G., Mack, C. L., Karpen, S. J., Hoofnagle, J. H., Doo, E., & Sokol, R. J. (2018). Biliary Atresia: Clinical and Research Challenges for the Twenty-First Century. Hepatology, 68(3), 1163-1173. PMID:29604222

Wang, L., Yang, Y., Chen, Y., & Zhan, J. (2018). Early differential diagnosis methods of biliary atresia: a meta-analysis. Pediatr Surg Int, 34(4), 363-380. PMID:29397405

