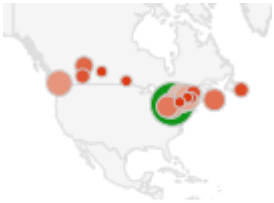
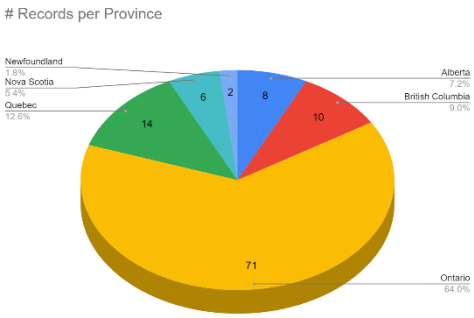
Canadian Biliary Atresia Registry | Registre canadien d'atrésie des voies biliaire

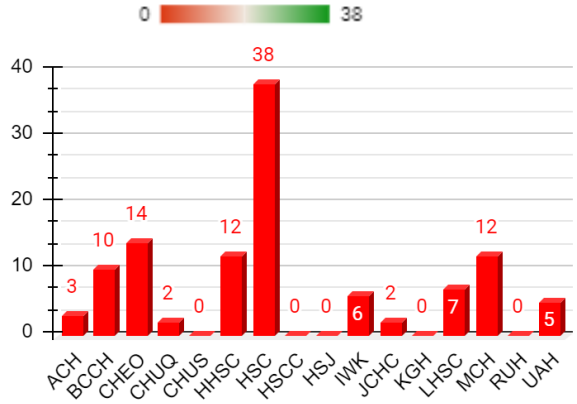
Newsletter - October 2022

Progress on CBAR Data

Nationally, we have collected data for 111 patients.







Website

The website will be updated over the course of the year and is now available in English and French.

Please send suggestions.

cbar.ca

Parent Committee

Elena is working on finding parents for the parent committee to increase involvement.

Discussions with families have frequently centered around early education and awareness for earlier diagnosis to improve Kasai success rates.

Please share interested contacts with Elena.

Upcoming Events to be scheduled – Stay tuned!

Steering Committee Meeting

National CBAR Strategic Planning Meeting

CBAR National Coordinator

If you have any questions or concerns about REDCap, REB, or other issues, please do not hesitate to contact Elena Guadagno at elena.guadagno@muhc.mcgill.ca

Child Foundation

La Fondation de l'Hôpital de Montréal pour enfants | The Montreal Children's Hospital Foundation

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Abstracts Presented

Thanks to Dr. Gil Eamer for preparing 2 abstracts.

G Eamer, RA Schreiber, S Emil, E Guadagno, D Briatico, BH Cameron and the Canadian Biliary Atresia Registry

CONTEMPORARY NATIONAL BILIARY ATRESIA INVESTIGATIONS AND OUTCOMES: A report from the Canadian Biliary Atresia Registry

Canadian Association of Paediatric Surgeons (CAPS) - Annual meeting 2021

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 pre-operative 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.

Table 1:
Diagnostic tests performed by age, concordance with biliary atresia diagnosis and days from referral to OR by test performed.

	Median age divided by if test was done						Mean days from referral to OR by tests done				
Pre-operative investigation performed	Test done Median age	n	Test not done Median age	n	p	% discordant with BA Dx	Test performed Mean days	n	Test not performed Mean days	n	p
HIDA	34	41	53.5	18	**0.02**	12%	14.0	41	12.7	18	0.66
Cholangiogram	57	19	37	39	0.35	21%	15.4	19	12.9	39	0.40
Liver biopsy	37	34	42	25	0.96	18%	16.8	34	9.2	25	**0.01**

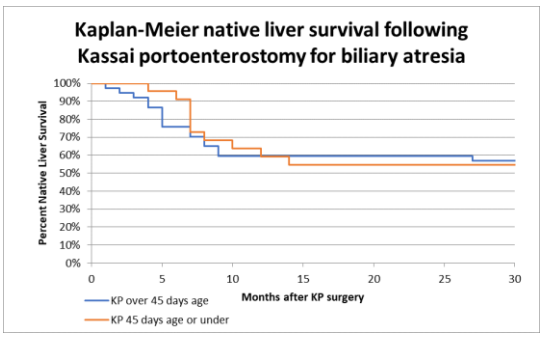
BILIARY ATRESIA NATIVE LIVER SURVIVAL FOLLOWING KASAI PORTOENTEROSTOMY: A report from the Canadian Biliary Atresia Registry

Pacific Association of Pediatric Surgeons (PAPS) - Annual meeting 2022

Purpose Multiple studies have found biliary atresia (BA) outcomes are improved when Kasai portoenterostomy (KP) is performed at a younger age. Multiple management algorithms have been developed to expedite time to KP through centralization and standardization. We have evaluated updated Canadian Biliary Atresia Registry (CBAR) data to assess time to surgery and native liver survival (NLS) following KP at CBAR centres.

Methods We analyzed prospectively collected CBAR data for pre-operative investigations, time to KP and long-term NLS with students t-test, chi-square analysis and Kaplan-Meier survival curves. The cohort was evaluated overall and by early (45-days and under) or late (over 45 days) age at KP.

Results Seventy-two patients presented to a CBAR centre with BA; median age at surgical consultation (42 days, n=65), days to surgery (10.5, n=64) and age at KP (53 days, n=69) were calculated. Time to surgery was significantly increased by liver biopsy (p=0.02), but not HIDA (p=0.52) or cholangiogram (p=0.38). Mean bilirubin change at discharge post-KP was -12 µmol/L and did not differ bdtween the early (3.1 µmol/L, n=18) and late (late -20.2 µmol/L, n=35) cohorts (p=0.06). NLS at 6 (n=58), 12 (n=54) and 24-months (n=52) was 83%, 59% and 54% respectively. Kaplan-Meier analysis (Figure 1) and chi-square analysis of age-at-KP's effect on NLS found no difference at 6 (p=0.07), 12 (p=0.18) or 24-months (p=0.13).



Conclusion Time to surgery is significantly delayed by percutaneous liver biopsy but not HIDA or cholangiogram. There is a trend towards longer NLS with early KP but wasn't not significantly different.

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Site Changes & Involvement

Site Name	Surgical SI	GI SI	Brief Status Update
Alberta Children's Hospital	Mary Brindle	Simon Lam replaces Steven Martin	Dr. Mary Brindle is adding Dr. Natalie Yanchar (Surgical) and Dr. Simon Lam (GI) who is replacing Dr. Steve Martin for CBAR.
Children's Hospital of British Columbia	Robert Baird	Rick Schreiber	Dr. Robert Baird (Surgical) and Dr. Rick Schreiber (GI) have recently hired a new coordinator, Deema Al, to help with local data.
Children's Hospital of Eastern Ontario	Ahmed Nasr	Carolina Jimenez	Dr. Ahmed Nasr (Surgical) and Dr. Carolina Jimenez (GI) continue to CBAR data collection. Dr. Manvinder Kaur was of great help in updating the data.
Centre hospitalier de l'université de Québec-Université Laval au CHUL	Julie Castilloux	Anna Wiexkowska	This site's data collection is on hold due to lack of resources.
McMaster Children's Hospital	Karen Bailey replaces Brian Cameron	Herbert Brill	Dr. Karen Bailey has replaced Dr. Cameron since his retirement. Dr. Brill continues to be the GI lead. Daniel Briatico is the coordinator assisting with data collection.
Hospital for Sick Children	Annie Fecteau	Simon Ling	Dr. Annie Fecteau (Surgical) and Dr. Simon Ling (GI) remain the site leads.
Children's Hospital of Winnipeg	Melanie Morris	Mark Deneau	Dr. Morris (Surgical) has been working on updating the local REB in hopes of collecting data in the near future with the help of Sylvia Froese, a regulatory coordinator.
Centre hospitalier Ste-Justine	Michel Lallier	Fernando Alvarez	We are awaiting a signature from Dr. Lallier (Surgical) for the contract and REB to be updated and Dr. Alvarez (GI) is available for data collection once everything is approved.
IWK Health Sciences Centre	Jessica Mills	Mohsin Rashid	Dr. Jessica Mills (Surgical) and Dr. Mohsin Rashid (GI) continue to work on data collection with the help of Bran MacIntyre, their local data collector.
Janeway Health Science Centre	David Price	Pushpa Sathya	Dr. David Price (Surgical) and Dr. Sathya (GI) continue to be the leads but no new data has been entered.
Kingston General Hospital	Andrea Winthrop	?	Dr. Andrea Winthrop (Surgical) is the lead but no patients have yet to be recruited.
Children's Hospital London Health Sciences Centre	Andreanna Bütter	Dandhapani Ashok	Dr. Andreana Bütter (Surgical) and Dr. Dandhapani Ashok (GI) continue to collect data with the help of Jacob Davidson, their local coordinator.
Montreal Children's Hospital	Jean-Martin Laberge & Sherif Emil	Najma Ahmed	Dr. Jean-Martin Laberge & Dr. Sherif Emil (Surgical) and Dr. Najam Ahmed (GI) continue with data collection with the assistance of Elena Guadagno.
Jim Pattison Children's Hospital	Amanda Hall	Garth Bruce	Dr. Amanda Hall (Surgical) is a new CBAR Site investigator and is diligently working on updating the REB and contracts in order for data collection to begin. Dr. Garth Bruce (GI) continues to be the lead GI.
Stollery Children's Hospital	*Bryan Dicken* replaces Gordon Lees	*Patricia Kawanda* replaces Jason Yap	Dr. Bryan Dicken (Surgical) has replaced Dr. Gordon Lees and Dr. Patrician Kawanda (GI) has replaced Dr. Jason Yap. Charlene Cars is the local coordinator who has helped with data collection. Cheri Copithorne (formerly Cheri Robert) has also been instrumental to get everything coordinated.