



THE ASSOCIATION FOR GLYCOGEN STORAGE DISEASE

The Ray

2017 Dues

The enclosed/attached “2nd Notice” membership form for 2017 dues is for those who have **NOT** paid for 2017. If you are not sure if you paid – call me (563-514-4022) and I’ll let you know. Remember, to be active members you must pay before March 1. Any dues (\$20) on or after March 1 will become \$15 for Associate dues/\$5 as a donation. NOTE: **We cannot receive your membership form by Facebook.** All monies received on Paypal with no membership form will be refunded. Please go to our website www.agsdus.org to complete the necessary form and payment or mail both to me: Hollie Swain, 611 Wisconsin Ave, Davenport, IA 52804. Thanking you in advance.

President’s Message

By Iris (Tremelling) Ferrecchia

A special thank you to Gus Paiva and Dr. Dwight Koeberl for the many years they served the AGSD Board. I will miss Dr. Koeberl’s straight forward opinion during board meetings and am grateful for the hours he spent preparing the GSD handbook. A big thank you and welcome to Jessica Knepler and Philippe Collat for committing time to take over the board positions of Gus and Dr. Koeberl.

Lots of gratitude is in order for many good things going on in the AGSD. Thank you to Kathy Thelen for continuing to work on the newsletter with Blair Stone-Schneider and Hollie Swain. Thank you to Brian Pawlik for keeping his hand on the communication and social media aspects of the organization. Thank you to Annalhees Elm and Irene Helley who are working with me to encourage more financial support from businesses. Thank you to Hollie Swain for her years of doing whatever job was needed, including keeping track of the organization’s finances.

At Dr. Dwight Koeberl’s suggestion, I contacted Lurie Children’s Hospital in Chicago to let them know about the AGSD conference in September. Professionals from the hospital, who have planned conferences such as ours, have volunteered to organize children’s sessions at the conference with the help of their strong Arts in Medicine program. I am grateful that they are so willing to participate with us.

If you have a desire to contribute your talents to the AGSD, please let me know. We can always use more hands to do that work which ultimately improves the quality of individuals affected by GSD.

2017 Annual AGSD Conference

By Jessica Knepler

I am honored to be hosting the 2017 Annual AGSD conference. Mark your calendars for September 15th and 16th and join us in Chicago! The conference will be held at the Hilton Garden Inn O'Hare in Des Plaines, IL. For convenience of those traveling by plane, there is a free shuttle from Chicago O'Hare Airport to the hotel. During your stay, there is also a shuttle from the hotel to the local train station, where you can hop a train to downtown Chicago to take in all the sights that our great town has to offer. Come early or stay after the conference to take in the Navy Pier, walk the Magnificent Mile, visit the Bean in Millennium Park the possibilities are endless! Keep watching the AGSD website (www.agsdus.org) for more information and details as they are approved. For those of you using Facebook, you can also join the FB event "AGSD 2017 Annual Conference".

The Annual GSD conference is an incredible time to not only learn from the great professionals that work tirelessly on research and treatments for GSD, but it is also an important time to come together with your GSD family and friends. I look forward to seeing you all in September!

Donations

It is with sincere appreciation that we gratefully acknowledge receipt of donations from the following people during the months of October, November and December, 2016.

General Fund

Vanguard Community Fund/
David Federico
YourCause
Mary & Emory Chapman
Brian & Sarah Narkewicz
Ken Edmonds
Dr. R Rodney Howell
Dan & Kathy Thelen
Network for Good/Michael Massei
Bill & Pat Tucker
Genzyme Corporation
Tilis Hofmann
Louis & Debra Becnel
Jewish Communal Fund
Matt LaGrow

In Honor of Jesse Narkewicz

David & Carol Gaylord

In Honor of Heather and Jonathan Edwards

Novel Insights Plus

In Memory of Jenniffer Kuttner

Sacramento Region Community
Foundation

Scholarship Fund

Ken Edmonds
Bill & Pat Tucker

In Memory of Rachel Walts

Kathy Thelen

In Memory of Brandon Bills

Kathy Thelen
Association for Glycogen Storage
Disease

In Memory of Dr. Barbara Brown

Anonymous
Dr. R. Rodney Howell
Hollie Swain
Dr. William Danforth

Research Fund

Joseph & Melissa Lipchick
Janice Schoen Henry
Mary Ellen Babich
Ken Edmonds
Margaret Baranoski
Bill & Pat Tucker
Nicholas Sabatine III
Jon & Becky Schumacher Wade
Teri & Fran Reed
Kathy Lupia

In Memory of Brian Tepperman

Association for Glycogen Storage
Disease

In Honor of Lucas Pond

Tower Center, LLC

In Honor of Justin Bagin

Carole Bagin

In Honor of Missy Lipchick

LLR, Inc

In Honor of Matthew & Brittany Byrne

Metanoia Properties LLC

Conference Fund

Esther James
Ken Edmonds
Bill & Pat Tucker
Genzyme Corporation
Louis & Debra Becnel
In Memory of Brian Tepperman
Kathy Thelen
In Honor of Jonah Helley & Family
Michael Myers

Family Assistance Fund

Ken Edmonds
Bill & Pat Tucker
Genzyme Corporation
Richard & Mary Jane Herrmann
In Memory of Iris Ferrecchia's Father
Kathy Thelen
In Memory of Dr. Ron Hofmann
Association for Glycogen Storage
Disease
In Memory of Ed Glover
Hollie Swain
In Memory of Ed Cronkleton
Hollie Swain
In Memory of Shirley Winegar
Hollie Swain

Synopsis of Minutes - AGSD Board of Directors Meeting Minutes Toronto, Ontario Canada, September 16, 2016

I. Call to Order

Chairperson Matt Peters called the Board Meeting to order at 10:38 a.m. ET. Also present were Iris Ferrecchia, Hollie Swain, Gwen Glover, Kathy Thelen, Gus Paiva, Richie Thelen, Laurie Kinnard, Dr. Dwight Koeberl and Brian Pawlik.

II. Approval of Meeting Minutes for May 23, 2016.

Minutes approved once corrections made.

III. Treasurer's Report

Financial Report for 2016 2nd Quarter was presented, discussed and approved.

Also new bank signing authorities were discussed and Hollie is to set those up accordingly.

IV. President's Report – for information only.

V. Vice-President's Report – for information only.

VI. Old Business

A. Updates on 2016 Conference Toronto, Ontario

Hotel contract, meal, conference registrations, sponsors, speakers, fundraising (Fun Run/Walk and Silent Auction), Saturday Night activity and parking expense discussed.

B. 2017 Conference proposal received from Jessica Knepler for Chicago, IL, discussed. September 15 and 16, 2017 at Hilton Garden Inn O'Hare Airport Hotel.

C. Two general board positions up for election; Gus Paiva and Dr. Koeberl's terms expire at end of the year. Hollie explained nomination process and ballot system. Gus Paiva nominated for position at this meeting and further nominations will be accepted at the General Meeting tonight.

D. Care Guidelines Update

- GSD types VI and IX guidelines have been submitted to Dr. M. Watson.

- Types V and VII: Hollie to check on progress of Type V and VII. Type IV – slated for 2017.

VII. New Business

A. SAB Update

Dr. Bachrach stepping down as SAB co-chair due to his busy schedule.

B. 2017 Proposal for Conference in Chicago, IL.

Preferred date September 15 and 16, 2017. Room rate \$119/night. Catering minimum \$5000 and then meeting room rental gratis. The hope is that professionals can convene at the conference to work on Guidelines for Type IV. Iris also expressed a desire to work with more professionals at Chicago Conference in order to offer more CME's.

C. 2018 - 40th Anniversary Conference Proposals

1. Proposal received from Iris for Radisson Resort at Gainesville, Florida.
2. Willing to entertain more proposals for 2018 and Hollie was asked to consider hosting in the Quad Cities for the momentous starting place of AGSD 40 years ago.
3. Possible 2018 conference dates of September 21 and 22, 2018 or October 12 and 13, 2018.

D. Scholarship Award

Tessa Peck recipient of \$1000 scholarship award.

E. AGSD Website, Facebook and Social Media in General

Brian indicated Website through Augie going well and he has set up various social media sites and experimenting with others. (e.g. hashtag for this conference, audio/visual equipment to record speakers, lavaliers attached to shirts, hand-held microphones, etc.)

Discussion regarding picture taking during conferences. (i.e. if permission granted to take pictures of individuals). Mike Venn, producer, creating a short film called "Me, My Friends, and GSD." Any participants at conference willing to be interviewed are to contact him at this time.

Mary was thanked for her part involving processing payments via PayPal; her tasks ended in October.

F. Membership Statistics

Hollie reported membership as 198 this year, an increase from 160 last year.

G. Conference arrangements: previously discussed.

H. Newsletter guidelines and submissions: There was a lengthy discussion as to what items can be included in the *Ray*. Item to be tabled for further discussion.

I. President's Notes

J. Other: None.

K. Next meeting: Tuesday, December 6, 2016 via conference call at 8:00 p.m. ET.

VIII. Adjournment

A motion to adjourn at 2:35 p.m. was approved.

Respectfully submitted,
Gwen Glover
Secretary.

Board Member Election Results

Thank you to the active AGSD members who were interested in running for an AGSD Board Member position in our most recent election. It was great to see several members wanting to be involved in our organization so that an election was needed to fill the two expired positions. Thank you also to all who took the time to vote and show your personal involvement in the AGSD.

We say welcome to Jessica Knepler and Philippe Collat as they each begin their three-year term; good-bye and thank you to Dr. Dwight Koeberl and Gus Paiva for their time and caring as retiring board members.

FYI to All Who Use VitaFlo Products

The AGSD was contacted by VitaFlo, one of our conference sponsors, about a new program that VitaFlo is offering to those who use their products. They asked if we could pass this information on to our members. The program, "VitaFlo4Success" is a program to help assist with insurance coverage for products that you might be using through VitaFlo. There is a short, but easy to follow tutorial about this new program if you wish to hear more information on this topic at vitaflo4success.com.

2016 End of Year AGSD Financial Report

STATEMENT OF CASH FLOW

INCOME

DUES	Active	\$2940.00	
	Lifetime	0.00	
	Associate	420.00	Total Dues 3360.00
DONATIONS	General	9995.02	
	Store	388.00	
	Scholarship	11,636.73	
	Research	9,999.00	
	Fund Raiser	0.00	
	Conference	12,565.27	
	Family Assistance	5,613.00	
	Pay Pal Acct	657.20	Total Donations 50,854.22
CONFERENCE	Registrations	13,143.00	
	Meals	120.00	
	Sat Meal (guests)	88.00	
	Fun/Run	4,418.50	Total Conference 17,769.50
INTEREST	Money Market Account	210.65	
	Savings Account	6.72	
	World Equities Acct	3,255.90	Total Interest 3,473.27
		TOTAL INCOME	75,456.99

EXPENSES

OFFICE	Telephone	0.00		
	Dues	200.00		
	Printing	0.00		
	Legal/CPA Fees/Insurance	932.45		
	Postage	286.43		
	Misc.	2,799.47		
	Bank/Finance Fees	0.00		
	Refunds	508.10		
	PayPal	551.63	Total Office 5,278.08	
	NEWSLETTER	Printing	1249.27	
		Postage	420.45	Total Newsletter 1,669.73
STORE	T-Shirts/bags/cards/etc.	0.00		
FUNDS	Conference			
	Rooms	259.86		
	A/V	2,495.09		
	Meals	12,254.05		
	Speakers	7,648.17		
	Sat. Night	3058.75		
	Fun/Run	730.34		
	Misc.	714.22	27,160.48	
	Research		0.00	
	Scholarship		1,000.00	
Family Assistance		646.10		
		TOTAL EXPENSES	35,754.39	

Decrease/Increase in Assets for Period Ended 12/31/16	39,702.60
Total Assets as of 12/31/16	172,139.45
TOTAL CASH HOLDINGS AS OF December 31, 2016	211,842.06

2016 End of Year AGSD Financial Report

	Beg Balance	Income	Expense	Transfers	Interest	Final Balance
General Fund	98,740.10	14,400.22	6,947.81	-3,000.00	1,806.34	104,998.85
Family Assistance	15,612.58	5,613.00	646.10	0.00	326.99	20,906.47
Research	35,576.62	9,999.00	0.00	0.00	724.74	46,300.36
Scholarship	1,806.36	11,636.73	1,000.00	0.00	34.09	12,477.18
Conference	-1,886.41	25,916.27	26,430.14	3,000.00	154.07	753.79
Fund Raiser	22,290.21	4,418.50	730.34	0.00	427.04	26,405.41
TOTAL	172,139.46	71,983.72	35,754.39	0.00	3,473.27	211,842.06

CASH IS HELD AS FOLLOWS:

Checking Account, Swain	65.77
Checking Account	17,877.00
Money Market Account	74,799.23
Savings Account	6,699.85
Capitol Securities	112,400.21
Pay Pal Account	0.00
TOTAL	211,842.06

GSD Ia and Kidney Disease

Monika Dambaska, MD

GSD I Kidney Disease is marked by enlarged kidneys, tubular dysfunction, glomerular (filter) dysfunction, proteinuria (protein excretion in urine) and kidney stones. The most common issue is glomerular hyperfiltration, which is similar to the kidney disease occurring in diabetes.

High blood pressure in GSD is most commonly related to kidney dysfunction. Increased pressure in the glomerulus (that serves as a filter) causes high blood pressure and protein excretion. ACE inhibitors (ACEi), such as Enalapril, Ramipril, Lisinopril, Losartan – Cozaar, Valsartan – Diovan are used to lower the pressure in the glomerulus (kidney) and help to prevent on-going damage.

A European Study on GSDI in 2002 claimed that 100% of GSD I patients had abnormal protein excretion by 24 years of age. In that same year Wolfsdorf and Weinstein proved that patients with kidney disease had worse metabolic control. An Italian study (2005) has shown slower progression in kidney function with the use of medications previously mentioned. Treatment with medications, however, did not prevent development of chronic kidney disease. In the Netherlands, Martens (2009) correlated optimal metabolic control with reno-protective effects on development of microalbuminuria. ACEi did not prevent progression of kidney disease, but none of the patients had good control. The next Italian study (2015) supported the importance of good metabolic control. Since complication in GSD has shown to

be preventable, we hypothesized that kidney disease could similarly be ameliorated if metabolic control improved in the patients taking ACE inhibitors. In our population, only 11.7% (23/195) of GSD Ia patients, and 13.3% (6/45) of GSD Ib patients developed abnormal protein excretion. It was most common in age group 16-20, which is consistent with other chronic conditions (lack of compliance in young adults). 11/14 patients with improved metabolic control and ACEi had diminished amount of protein in their urine. Three participants who worsened while on ACE inhibition had no change in their metabolic control, but no patient developed chronic kidney disease. Dietary compliance with strict metabolic control is likely to prevent the onset of kidney disease. Metabolic control plus medication for the patients with established GSD Ia glomerulopathy is necessary and progression of glomerulopathy can be reversed.

During pregnancy, ACEi must be discontinued, but no long-term worsening has occurred in any pregnancies followed by our institution (49 deliveries). The GSD population is also very prone to kidney stones, which is not correlated with metabolic control. Citrate supplementation prevents this complication. Kidney disease was not detected in any of the 135 patients with the ketotic form of GSD (types 0, III, VI, IX) even though they are on very high protein diet. Liver transplant does not prevent GSD I kidney disease, and anti-rejection medications are toxic to the kidneys.

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