SUMMER 2014

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Les Guédry d'Asteur

IN THIS ISSUE

With our 7th major reunion of the Guédry and Petitpas families just six weeks distant we are in the final stages of planning. It will be a great time for all Guédry and Petitpas cousins to gather, renew old acquaintances and make new friends. We have an outstanding agenda with the Louisiana Cajun band "Chér Mom" featuring the Christine Guidry Law family, a rare French Mass, interesting presentations, tasty Acadian food and, most importantly, lots of time to mingle and meet with cousins. We'll also have displays about our families and family-related items to purchase. Review the Agenda in this issue to learn more about the Reunion on 16 August 2014 at the Acadian Village just north of Van Buren, Maine. In the Spring 2014 issue of "GENERATIONS" we published detailed information on accommodations, transportation available to Van Buren and other traveling facts. The Summer 2013 issue of "Generations" contained a travelogue of Acadian sites to visit in the Madawaska region while attending the Reunion and Congrès Mondial Acadienne 2014. You can view these two issues at these links:

http://freepages.genealogy.rootsweb.ancestry.com/~guedrylabinefamily/spring2014newsletter.pdf (Spring 2014 Issue)

http://freepages.genealogy.rootsweb.ancestry.com/~guedrylabinefamily/summer2013newsletter.pdf (Summer 2013 Issue)

We will have some souvenir items for sale at the Reunion, but will not have shirts and other clothing items. We have arranged, however, to have these types of items available for purchase on CafePress.com with our 2014 Reunion logo. Just go to this link and select what items you would like to purchase – then wear them at the Reunion.

http://www.cafepress.com/+guedry-1014+gifts

In this issue we have an in-depth article on major Acadian Genetic Diseases that should be of interest to anyone with Acadian ancestry. These diseases are not exclusive to the Acadian population, but occur at a greater frequency among Acadians than the general population. Many can be traced back to a single individual (by name) living in Acadia during the seventeenth century. Also, read about our featured Family Musician, Emilie Guidry, currently playing at Pat O'Brien's in the New Orleans French Quarter. Want to try some truly Maine Acadian dishes before the Reunion or prepare your palate for your visit to the Madawaska area? Then flip to the Bon Appetit page of "GENERATIONS" and enjoy some Ployes for breakfast and a bowl of unique Soupe aux Pois (Acadian Pea Soup) for dinner. Our Book Nook in this issue contains two interesting books on the history of the Aroostook, Maine area and on traditional Maine Acadian cooking. And my favorite section of the newsletter – Historical Tidbits – contains several interesting articles from newspapers of yore about our families.

As a final note, Brittany Guidry of Houma, Louisiana – the reigning Miss Louisiana USA 2014 – was third runnerup in the Miss USA 2014 contest held in early June in Baton Rouge, LA. Brittany was an outstanding representative of Louisiana, Acadians worldwide and the Guédry family in particular.

ACADIAN GENETIC DISEASES

by Martin Guidry

THE INSULAR ACADIANS

Acadians experience a high incidence of certain genetic diseases compared to the North American population as a whole. This is not uncommon among relatively isolated ethnic groups. Because of the importance of this topic to Acadians everywhere, we'll briefly discuss aspects of Acadian history that relate to this high incidence, a review of genetics and disease and finally genetic diseases with an Acadian component.

It is important to understand that DNA (deoxyribonucleic acid) research in studying genetic diseases is not related to the DNA analyses used in genealogical research. Different segments of the chromosome are used in medical research and in genealogical testing; therefore, the information obtained is unrelated.

The Acadian population prior to the deportations of 1755-1764 descended from a relatively small number of original settlers (several hundred at most) that came primarily from France. In Acadia they settled in several relatively isolated communities among their extended families (i.e., parents, uncles, aunts, siblings, cousins). To survive, the deported Acadians again grouped closely together and made every effort to find and join their extended families. This was true both in their initial settlements prior to 1763 in the Atlantic seaboard colonies, France, England and eastern Canada as well as their resettlements in Louisiana, Maine, New Brunswick, Nova Scotia and Québec.

Historically, the tenets of the Acadian people have been Catholicism, family, French language and their Acadian culture. These are strong yet today in Acadian regions throughout North America with Acadians clustered close together in their villages and towns of Louisiana, Maine, New Brunswick, Nova Scotia and Québec.

The historic, insular nature of the original Acadian society resulted in a common genetic inheritance. Having maintained their familial and social cohesiveness during the ensuing 350 years has resulted in an increased risk among Acadians for rare genetic diseases. Although intermarriage among closely-related relatives has occurred in the Acadian population, this is not the major factor in explaining the high incidence of genetic diseases among the Acadians. The small pool of Acadians settling in isolated communities, maintaining their unique culture and seldom marrying outside of the community led to a small gene pool with a high disposition for genetic diseases.

GENETICS AND MUTATIONS

All humans are remarkably similar – 99.9% similar even though we have obvious physical differences as hair color, skin color, eye color, height, weight, etc. This similarity is due to the chemical DNA found in all humans. A long and complex chemical, DNA is instrumental in cellular reproduction in the body. Each time the body must make a new cell, the original cell must undergo over three billion chemical reactions to make a new copy of its DNA for the new cell. It is essential that the original cell reproduce its DNA as perfectly as possible. Humans are composed of trillions of cells that live, die and often are replaced over the course of a lifetime thus these billions of chemical reactions to reproduce the DNA in each new cell must occur perfectly innumerable times over a lifetime.

DNA serves as the source of information for cell structure and function. The DNA-encoded information directs the cell to produce other chemicals called proteins needed for the various cell functions. Proteins control the physical attributes as eye, hair and skin color, height, weight, etc. By making a perfect copy of its DNA, each cell ensures that the protein it produces will contribute positively to the health of the human being. The DNA segment that makes a specific protein is called the gene. Each human has approximately 35,000 genes.

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Each human baby results from the union of the sperm cell of his father and the egg cell of his mother. In this way DNA is transmitted from the parent's cells to the baby's cells. Each baby receives two copies of DNA – one from each parent; therefore, the cells of the baby will be making the proteins of his parents. Ever wonder why we look similar to our parents?

Babies, however, are not identical to their parents nor are any two human beings identical. Although cells attempt to reproduce their DNA perfectly, occasionally mistakes are made. These errors are quite rare and seldom affect genes or their ability to encode proteins. Very rarely, however, one of these errors does affect the protein produced and its function. These "changed" genes that affect the proteins are called mutations and they may cause medical problems as well as alter physical attributes as eye color.

Should the mutation occur in the sperm cells or egg cells of the parents, the mutation could be transmitted to their babies. If transmitted to a baby, it will occur in the DNA of all cells of the baby as it matures to adulthood because all adult cells derive from the original sperm cell and egg cell received from the baby's parents. Thus the mutation can be transmitted to consecutive generations. When a mutation causes a medical problem seen in a family across several generations, scientists refer to the medical problem as a genetic disease.

Humans receive two copies of DNA from their parents – one from the sperm cell and one from the egg cell. Thus humans have two copies of each of the approximately 35,000 genes. Genetic diseases can be either dominant or recessive. In a dominant genetic disease only one mutated gene of the pair is needed to cause the disease. In a recessive genetic disease both genes of the pair must be mutated before the disease is observed. For a recessive genetic disease having one mutated gene and one normal gene does not result in the genetic disease. Most Acadian genetic diseases are recessive genetic diseases.

In summary, each person has two copies of each gene on a chromosome and in almost every instance both copies of each gene are normal. Rarely are one or both copies of a gene on the chromosome mutated. Each parent transmits one copy of each of his or her genes to their baby. The baby thus receives two copies of each gene – one from the father and one from the mother.

For a specific gene causing a dominant genetic disease, the baby must receive a mutated gene from at least one parent in order to have the disease. If one parent has two mutated genes, all of the children will have the disease. If each parent has one mutated gene, there is a 75% chance that each of their children will have the disease. If one parent has one mutated gene and the other parent has two normal genes, there is a 50% chance that each of their children will have the disease.

For a specific gene causing a recessive genetic disease, the baby must receive a mutated gene from each parent in order for the baby to have the genetic disease. If the baby receives one mutated gene and one normal gene, then the baby will not have the genetic disease, but will be a carrier of the disease and could transmit the genetic disease to his children. For a baby to receive two mutated genes of a recessive genetic disease, each parent either must have the genetic disease or be a carrier of the genetic disease (i.e., either one or both of each parent's genes must be mutated). If both parents are carriers of the genetic disease, there is 25% chance that each of their children will have the genetic disease. If one parent is a carrier and one parent has the genetic disease, there is a 50% chance that each of their children will have the genetic disease. If both parents have the genetic disease, all of their children will have the genetic disease.

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Often an Acadian genetic disease can be traced back to a specific couple living in seventeenth century Acadia. Assume that the husband of this couple accidentally developed one mutated gene of the gene pair. The wife had two normal copies of this gene. There is a 50% chance that any of the children of this couple would inherit the mutated gene. Assuming this gene is recessive, then any children that inherit the mutated gene would be carriers of the genetic disease and would not have the genetic disease. If the couple had ten children, which was not unusual for an Acadian couple of that time, then statistically five of the children would be carriers of the genetic disease. If these five children each had ten children with spouses that did not have the mutated gene, then statistically 25 more children would be carriers of the genetic disease. These could be both boys and girls. Continuing in the same manner for two more generations would result in an additional 125 carriers of the genetic disease in the fourth generation (2nd cousins) and 625 carriers in the fifth generation (3rd cousins). Thus in a relative few generations several thousand distant relatives would be carriers. Within the isolated Acadian communities comprising extended families, it is probable that two distant relatives (e.g., 5th cousins) who are both carriers of the genetic disease marry. Their children would each have a 25% probability of inheriting two mutated genes (i.e., one from each parent) and thus having the recessive genetic disease. Recessive genetic diseases, therefore, do not have to result from close relatives having children, but can "jump" several generations before appearing in the population.

THE ACADIAN DISEASES

The genetic diseases often called Acadian diseases were not exclusive to Acadians, but occur within several different groups in North America. They, however, occur at a higher frequency among the Acadians than the North American average.

The major Acadian genetic diseases are:

* Acadian Usher Syndrome (Type Ic)
* Tay-Sachs Disease
* Acadian Ataxia (Friedreich Ataxia)
* Charcot-Marie-Tooth Disease (Type IA)
* French Settlement Disease (Hereditary Spastic Paraplegias)
* Niemann-Pick Disease (Type C2)
* Acadian Variant Fanconi's Syndrome

Acadian Usher Syndrome

Discovered among the Acadians in 1996 by Dr. H. W. Kloepofer when doing research at the Louisiana State School for the Deaf in Baton Rouge, Usher Syndrome impacts young children at birth. They are born deaf and eventually begin losing their eyesight during adolescence.

Usher Syndrome accounts for the majority of persons in the United States who are both deaf and blind. Usher Syndrome is an autosomal recessive genetic disease. (Autosomal means the affected gene is on any chromosome except one of the two sex chromosomes.). Approximately 4 persons of every 100,000 Americans have Usher Syndrome although about 1 in 100 Americans carry one Usher Syndrome defective gene and, therefore, are carriers of the disease. There are three clinical types of Usher Syndrome (Type I, Type II and Type III) with six subtypes of Type I. Ten different genes when mutated could cause Usher Syndrome. Type I is the most severe form of Usher Syndrome.

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Acadian Usher Syndrome is Type Ic characterized by congenital deafness and progressive blindness. In 2000 scientists discovered the mutated gene, *harmonin*, that causes Type Ic Usher Syndrome. Unlike the normal harmonin gene, the mutated harmonin gene cannot produce the protein harmonin needed for hearing and vision. This disease is quite rare among the Acadians in Louisiana with only about 300 confirmed Acadian patients since 1966 among the approximately 1,000,000 Acadians in Louisiana. The frequency of Acadian Usher Syndrome is 1 in 20,000 Acadians in Louisiana today with 1 in 70 Louisiana Acadians being a carrier. Because Acadian Usher Syndrome is a recessive genetic disease, the affected patient must have inherited one mutated haromnin gene from each parent; therefore, this patient would have no genes that can produce the protein harmonin.

The protein harmonin is important in maintaining the structure and function of cells that comprise the inner ear and the retina. The inner ear receives and transforms vibrations into impulses that the brain interprets as sound. Since Acadian Usher Syndrome patients have no harmonin protein, their inner ear does not transmit impulses to the brain. Their hearing is so severely impacted that hearing aids are of no benefit and only cochlear implants offer hope to the patient. The retina is located in the rear of the eye. It absorbs light and transmits it as impulses that the brain interprets as vision. At birth the retina is fine; however, over time the retina does not receive harmonin needed to maintain it and it slowly begins to degenerate. As the child nears ten years of age night blindness begins and the field of vision decreases. The loss of vision gradually becomes more severe until total blindness occurs in early adulthood.

Identifying the mutated gene was a major breakthrough in understanding Acadian Usher Syndrome and helping affected patients. A blood test has been developed that offers pre-natal diagnosis for Acadian Usher Disease and also can identify carriers of the disease. Hearing-impaired infants can be tested and, if affected by Acadian Usher Syndrome, can undergo therapy programs with their parents. Genetic counseling can be provided to carriers to assist them in their decisions. Researchers currently are studying the potential of cellular therapy to prevent or reduce blindness in Acadian Usher Syndrome patients.

DNA research on Type Ic Acadian Usher Syndrome patients strongly suggests that the origin of this disease was an accidental mutation in a common ancestor living in Acadia in the seventeenth century. Following the Acadian deportations from 1755 – 1764 approximately 3000 Acadians eventually settled in small, close-knit, familial Acadian communities in southern Louisiana. Essentially all carriers of Acadian Usher Syndrome arrived in southern Louisiana in 1766 and descended from the individual above with the mutated gene. They settled in only a few, small, isolated Acadian communities with extended families that historically have not moved far from the parental home. Marriages in these isolated communities often occurred between distantly related individuals who were both carriers of Acadian Usher Syndrome. This has resulted in Acadian Usher Syndrome being found primarily in three parishes of southwestern Louisiana – Lafayette, Vermilion and Acadia.

Tay-Sachs Disease

Tay-Sachs is a fatal, autosomal recessive genetic disease with no known cure. Shortly after birth, infants become paralyzed over their entire body as the disease attacks their nervous system. Untreatable, Tay-Sachs successively causes loss of motor skills, seizures, blindness, deafness, paralysis and finally death by the age of five. Tay-Sachs is not clinically apparent until about six months of age although the disease begins its destruction in the fetus early in the pregnancy.

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In south Louisiana Tay-Sachs is known as "the Cajun disease". "Lazy Baby Disease", a similar affliction of the past in Louisiana, likely was undiagnosed Tay-Sachs Disease.

Generally associated with the East European Ashkenazi Jews, Tay-Sachs Disease is rare among the Acadians of south Louisiana. The congenital absence of the enzyme Hexosaminidase-A causes Tay-Sachs Disease. Without this vital enzyme the body cannot break down one of its fatty substances, the lipid ganglioside GM2, causing this lipid to build up in the nerve cells of the brain and impair the central nervous system.

Tay-Sachs Disease in Louisiana is greatest in four southwestern Louisiana parishes – Allen, Acadia, Jefferson Davis and Lafayette. As expected, the carrier frequency is also much higher in the affected parishes. For example, a study in Allen Parish found that the Tay-Sachs carrier frequency is ten times greater than that of the general population. Genealogical and DNA studies have shown the disease arrived in southwest Louisiana before 1850 and probably when the Acadians first settled in these Louisiana communities. A single ancestral couple has been identified that is common to almost all of the known Acadian Tay-Sachs patients.

A tell-tale sign of Tay-Sachs Disease is a cherry-red spot in the eyes of the patient. Today a blood test can determine the level of the enzyme Hexosaminidase-A in the fetus or infant and thus the presence of Tay-Sachs Disease. Also, testing of serum and white blood cells can confirm if a person is a carrier of Tay-Sachs Disease. Affected persons can then receive genetic counseling to assist them in dealing with the patient and with their decisions as carriers.

Encouragingly, researchers seeking a cure for Tay-Sachs Disease have discovered a drug that blocks the accumulation of fatty substances in the brain. This drug, a small, artificial imino sugar molecule, blocks the first step in the synthesis of the types of lipids that accumulate in the brain of Tay-Sachs patients. If the production of these lipids can be stopped, then the devastating effects of the disease will not occur. Other therapies being researched by scientists include gene therapy, neural stem cells, bone marrow transplants and metabolic bypass therapies.

Acadian Ataxia (Friedreich Ataxia)

Acadian Ataxia, a form of Friedreich Ataxia, is found in individuals of Acadian ancestry in Louisiana and eastern Canada (Nova Scotia, New Brunswick and eastern Québec). Acadian Ataxia is a progressive neurodegenerative disease involving both the central and peripheral nervous systems. It is an autosomal recessive genetic disease that destroys brains cells governing muscle control. Usually appearing about puberty, Acadian Ataxia first appears as slurred speech, a stumbling walk and hand incoordination followed by loss of ability to walk, stand and move. There is no known cure for Acadian Ataxia.

Compared to other forms of Friedreich Ataxia, Acadian Ataxia has a slower progression and less severe secondary symptoms as hearing impairment. The age of onset of Acadian Ataxia is slightly later and the age of death older.

A form of muscular dystrophy, Acadian Ataxia can be mistaken for multiple sclerosis and other diseases affecting coordination.

In the U. S population one in 50,000 people inherit Friedreich Ataxia while one in 122 people are carriers. In southwest Louisiana one in 20,000 Acadians inherit Acadian Ataxia and one in 70 are carriers. As with other

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Acadian genetic diseases, Acadian Ataxia is most prevalent in a few small Acadian communities of south Louisiana, Nova Scotia, New Brunswick and eastern Québec.

Scientists have determined that the Acadian Ataxia defective gene is on chromosome 9 of the 46 chromosomes that humans have. Researchers are working diligently to identify the defective gene and the protein it produces.

Blood tests currently can identify individuals with Acadian Ataxia as well as carriers of the disease. Although there is no known cure for Acadian Ataxia, patients can receive counseling and therapies, including physical aids and medical intervention, that relieve the symptoms.

Charcot-Marie-Tooth Disease

A form of ataxia, Charcot-Marie-Tooth Disease (CMT) is a progressive neurological disease that leads to deterioration of muscles in the feet, lower legs, hands and forearms. In CMT the peripheral nerves, those outside the brain and spinal cord, are affected. The first symptom of CMT is normally a foot deformity as high arches or flexed toes which affect walking and causing tripping, raising the feet higher than normal, walking with a gaited step and numbness in the feet. The lower arms and hands may later be affected – sometimes so severely that the hands cannot be used.

Dr. Carlos Garcia discovered CMT among the Acadians of south Louisiana in 1970. There are four types of Charcot-Marie-Tooth Disease (Types I-IV) and seven subtypes of Type I. The Acadian form of Charcot-Marie-Tooth Disease is Type IA. Unlike other Acadian genetic diseases, Charcot-Marie-Tooth Disease is an autosomal dominant genetic disease thus a person need only receive one defective gene from his parents to acquire the disease. At least one of his parents will have CMT Type IA.

A duplication of the PMP22 gene on chromosome 17 causes CMT Type IA. Instead of having two copies of the PMP22 gene (i.e., one on each paired chromosome), CMT Type IA patients have three copies of the gene (i.e., two on one chromosome and one on the paired chromosome). The PMP22 gene produces peripheral myelin protein. The exact function of this protein in causing Charcot-Marie-Tooth Disease is not known. Similar to most other CMT patients, Type IA patients initially are slow runners in childhood, develop high arches, hammertoes and weak ankles affecting walking. Hand weakness usually occurs about ten years after foot and leg problems. These patients may develop hearing and vision deficiencies. Generally patients remain ambulatory throughout life with a normal life expectancy.

Blood tests can determine if a person has Charcot-Marie-Tooth Disease. Historically, electrodiagnostic testing as nerve conduction velocity tests and electromyograms have been used. Patients with CMT can receive physical and medical assistance such as specialized shoes, wearing braces or using a wheelchair for mobility and undergoing surgery to improve the foot, leg and hand structure. Physical therapy and genetic counseling are also important. In southwestern Louisiana CMT seems to be the most prevalent form of muscular dystrophy.

French Settlement Disease (Hereditary Spastic Paraplegias)

French Settlement Disease (FSD) is a rare genetic disease first discovered in French Settlement – a small village in Livingston Parish, LA. It is an autosomal recessive genetic disease and a form of genetic diseases called Hereditary Spastic Paraplegias (HSP).

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Hereditary Spastic Paraplegias affects the muscles and movements of the lower limbs and torso of the body. The main symptom of HSP is difficulty walking due to weak and stiff leg muscles which begins suddenly and worsens with age. It often becomes quite severe and debilitating. The first symptoms are often balance issues, stumbling and stubbing the toe. Occasionally HSP can affect the upper body as the arms and can cause problems with speech and swallowing.

A deterioration of specific nerve cells (i.e., the upper motor neurons) in the brain and spinal cord that control movement of voluntary muscles causes Hereditary Spastic Paraplegias. When the circuit between the upper motor neurons and the muscles is broken due to this deterioration, then control of voluntary movements is lost. Deterioration of the upper motor neurons occurs because a mutated gene cannot produce the proper protein essential for the maintenance of the neurons.

French Settlement is a small community of approximately 1000 persons along the Amite River in Livingston Parish. In the 1700s and early 1800s several cultural groups settled in the French Settlement area including Acadians, Germans, Spanish, French and Italians. Acadians are the most prominent ethnic group in the community. French Settlement Disease was first identified in this population in 1976. Genealogical research of the FSD patients revealed that French Settlement Disease originated from a specific German couple who immigrated to the French Settlement area in 1741.

The differences between French Settlement Disease and other forms of Hereditary Spastic Paraplegias are difficult to distinguish. For FSD the age of onset is typically in the mid-twenties whereas other HSP forms first appear at one to thirteen years of age. Most physical symptoms of FSD and other HSP forms are similar.

The identity of the mutated gene causing French Settlement Disease is not known yet; therefore, no genetic test is available to diagnose this disease before onset of symptoms. Having a genetic test available would allow individuals having FSD to begin in youth a lifestyle that would strengthen the limbs. A genetic test could also identify carriers of FSD and thus provide them with genetic counseling.

Recently an Acadian from northeastern Maine who grew up in a French Acadian community was diagnosed with Hereditary Spastic Paraplegias. Neither the type of HSP nor the cause of this person's HSP is known presently.

Niemann-Pick Disease

Niemann-Pick Disease (NPD) is an autosomal recessive genetic disorder resulting from an accumulation of fat and cholestrerol in the cells of the liver, spleen, bone marrow, lungs and occasionally brain. Physical symptoms of Niemann-Pick Disease are loss of muscle control, eye paralysis, brain degeneration, learning problems, spasticity, swallowing difficulties, slurred speech, hypersensitivity to touch and corneal clouding. About half of the Niemann-Pick patients develop a cherry-red halo around the center of the retina. There is no known cure for Niemann-Pick Disease.

There are four categories of Niemann-Pick Disease (Types A, B, C1 and C2) with Type A being the most severe form. Originally, Types C1 and C2 were known as Types C and D. Most Niemann-Pick Disease Type C2 patients are Acadians of western Nova Scotia. In these patients the disease usually develops in school-age children although occasionally symptoms develop in the adult years. The disease progresses slowly.

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A mutation of the NPC2 gene causes Niemann-Pick Disease Type C2. The NPC2 gene provides instructions to produce a protein that binds and transports cholesterol. A lack of this NPC2 protein leads to an abnormal accumulation of lipids and cholesterol in the cells.

In Type C2 patients lipids and cholesterol accumulate within the liver and spleen and excessive amounts of other lipids build up in the brain. Type C2 patients have moderately large spleens and livers, an inability to look up and down, difficulty in walking and swallowing, progressive loss of vision and hearing and seizures. Life expectancy varies considerably with many dying in their teens while some live to forty years of age.

A genetic test has been developed that can diagnose if a person has Niemann-Pick Disease Type C2 and also if a person is a carrier of the disease. Genetic counseling can then assist carriers in making needed decisions. NPD Type C2 patients can obtain physical and medical therapy such as devices to assist in walking, hearing and vision as well as low cholesterol drugs and medication for seizures.

The frequency of Niemann-Pick Disease Type C2 is 1 in 150,000 people and usually occurs in Acadians of western Nova Scotia – particularly in Acadians originating from Yarmouth County where from 10% to 26% of the population are carriers. Genealogical studies indicate Niemann-Pick Disease Type C2 originated from the Acadian couple Joseph Mius dit d'Azy and Marie Amirault in the late seventeenth century.

Acadian Variant Fanconi's Syndrome

Fanconi's Syndrome is a disorder of the proximal renal tubules of the kidney in which substances as glucose, amino acids, uric acid, phosphate and bicarbonate are passed into the urine rather than being reabsorbed.

Acadian Variant Fanconi's Syndrome initially was identified in 1971 among Acadian children in Maritime Canada. The Acadian Variant is observed initially in young children between three and ten years of age most commonly suffering from "knocked knees" and growth failure as well as rickets and some degree of renal insufficiency. Renal insufficiency progresses slowly and often deteriorates during adolescence into chronic kidney disease.

Childhood therapy includes phosphate supplements, alkali replacement and vitamin D resulting in improvement of rickets and leg alignment. If renal failure occurs, patients may have to undergo kidney transplantation. Acadian Variant Fanconi's Syndrome is an autosomal recessive genetic disease.

DALHOUSIE UNIVERSITY
HALIFAX, NOVA SCOTIA, CANADA

LSU HEALTH SCIENCES CENTER
NEW ORLEANS, LA

TULANE UNIVERSITY MEDICAL CENTER
NEW ORLEANS, LA

ACADIAN GENETIC DISEASES

by Martin Guidry

THE RESEARCH FACILITIES

Several medical laboratories and facilities within North America are conducting research on Acadian genetic diseases. Below is a brief list of research facilities studying Acadian genetic diseases and some of the researchers working on these diseases. This list is not comprehensive.

* Louisiana State University Health Sciences Center – Center for Acadiana Genetics and Hereditary Health Care (New Orleans, LA)
  - Dr. Bronya J. B. Keats, Ph. D.
  - Dr. Yves Lacassie, M.D.
  - Dr. Michal Jazwinski, Ph.D.
  - Dr. Sevtap Savas, Ph.D.
  - Dr. Mary Z. Pelias, Ph.D.
  - Judy LaBorde

* Tulane University Medical Center Human Genetics Program (New Orleans, LA)
  - Dr. Jess Thoene, M.D.
  - Dr. Carols Garcia, M.D.
  - Dr. Hans Andersson, M.D.

* Children's Hospital (New Orleans, LA)
  - Dr. Alan Robson, M.D.

* Nicholls State University (Thibodaux, LA)
  - Dr. John Doucet, Ph.D.

* Louisiana State University (Baton Rouge, LA)
  - Dr. Mark Batzer, Ph.D.

* McGill University (Montréal, Québec, Canada)
  - Dr. Charles Scriver, M.D.

* Dalhousie University Medical School (Halifax, Nova Scotia, Canada)
  - Dr. Peter R. Camfield, M.D.
  - Dr. Wenda Greer, Ph.D.
  - Dr. D. Christie Riddell, Ph.D.
  - Dr. David M. Byers
  - Dr. Paul E. Neumann, M.D.

* Boston College (Chestnut Hill, MA)
  - Dr. Thomas Seyfried, Ph.D.

* Baylor College of Medicine (Houston, TX)
  - Dr. James R. Lupski, M.D.
  - Dr. Philip F. Chance, M.D.

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An excellent website with information on Acadian Genetic Diseases is "Genetics and Louisiana Families" at: http://www.medschool.lsuhsc.edu/genetics_center/louisiana/

Recently Dr. John Doucet, Professor of Molecular Genetics at Nicholls State University, expressed the importance of Acadian families in research on genetic diseases: "Given our history of exile and displacement and isolation, generations of Acadian descendants should have little reason to trust other peoples. Yet we do. And, importantly, we trust genetic scientists. And by trusting geneticists, we help people around the world understand those same genetic disorders that run in our own families. For this reason alone, I think that Acadians are the most selfless people on earth."

Dr. John Doucet
Professor of Molecular Genetics
Nicholls State University

Baylor College of Medicine
Houston, TX

Dr. Bronya Keats
LSU Health Sciences Center
New Orleans, LA

CONGRÈS MONDIAL ACADIEN 2014

L'ACADIE DU MONDE

The Guédry & Petitpas Family Reunion

16 August 2014

Van Buren, Maine

9:00 AM - 5:00 PM

Acadian Village

BON APPETIT

PLOYES (BUCKWHEAT PANCAKES) AN ACADIAN BREAKFAST STAPLE-
Bouchard Family Farms Cookbook
(*see Book Nook*)

1 cup yellow buckwheat flour
1 cup regular flour
4 tsp. baking powder
1 tsp. salt
1-1/2 cups cold water
1/2 cup boiling water

Mix dry ingredients. Add cold water and let stand for 10 minutes. Add boiling water and drop to make thin 6" pancakes on hot griddle, 400 degrees (200 C.) Bake on one side only, until bubbled and firm. Serve on warm platter, cover with napkins.

Buckwheat pancakes are a venerable old mainstay of French cookery and one finds variants of them in many parts of French North America, including the St. John Valley, the Acadian Maritimes, and Quebec.

ACADIAN PEA SOUP (Soupe aux pois)
Bouchard Family Farms Cookbook
(*see Book Nook*)

1 lb. whole dried yellow peas
8 cups water
1/2 lb. salt pork (or a ham bone)
1 large onion, chopped
1/2 cup celery, chopped finely
1/4 cup carrot, grated or chopped finely
1 tsp. dried savory

Wash and sort peas; place in a large pot, bring to a boil. Remove from heat and let sit for 1 hour. Add salt pork, onion, celery, carrots, savory and 1 tsp salt. Bring to a boil; reduce heat and simmer until peas are very tender, about 2 hours, adding more water if needed. Remove salt pork; chop and return to soup. Season to taste with salt and pepper.

RUTH GUIDRY'S BUTTER CREAM FROSTING
(Charlene Guidry Lacombe-Jennings, LA)

1 cup milk
3 tbs. all-purpose flour
1 cup sugar
1/2 cup butter
1/2 cup margarine
1 tsp. vanilla

Combine milk and flour, stir and cook until thick. Let cool. Mix the other ingredients and beat until fluffy. Combine mixtures and beat well.

FAMILY MUSICIANS
EMELIE GUIDRY

Born in 1984 in Lafayette, Louisiana, Emelie Guidry is a singer, songwriter, pianist and guitarist. Emelie's voice lights up a room. She has a blend of optimism, idealism and quirkiness. *Groovescapes New Orleans* describes Emelie as "a talented songwriter with a captivating stage presence" and her songs have been described as "musically adventurous... an impressive and unexpected blend" by the *Times of Acadiana*.

The daughter of two music lovers, Emelie's childhood soundtrack was an eclectic mix of oldies radio, 80's-90's hits and the immersion of zydeco, second-line, jazz and blues. Adding her Cajun culture into this mix gave Emelie that unique style that draws crowds to her. Teaching herself piano from age 4, learning clarinet at 10 and guitar at 15 led Emelie to a love of songwriting, singing and eventually in 2007 a Bachelor's Degree in Music Theory and Composition from the University of Louisiana at Lafayette.

Emelie has performed throughout the south as both a solo performer and with many cover bands, choirs, jazz ensembles and her own group, The Picardy Birds. Sharing her vigor and matching her dynamism, the band members draw out the tension between Emelie's lyrics and verses using sparkling grooves, winding psychedelic stretches and syncopated jazz flourishes. The Picardy Birds, led by Emelie as vocalist and acoustic guitarist, released their first full-length album, **Play On**, in 2008.

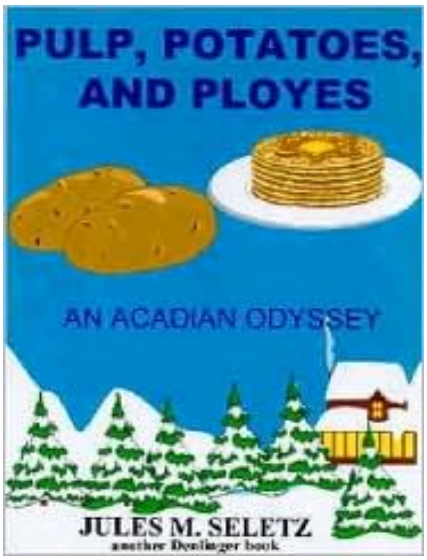
In 2012 Emelie made a career move to New Orleans where she is an entertainer at Pat O'Brien's Piano Bar. Currently playing four to five nights a week, she draws large crowds to listen to her special style of music. Emelie recently released a self-titled EP and has begun work on a full-length album of collaborations with various Louisiana musicians.

You can listen to Emelie Guidry at: www.facebook.com/emelieguidrymusic and soon at her website emelieguidry.com .

Emelie Guidry at Pat O'Brien's

BOOK NOOK

PULP, POTATOES, AND PLOYES AN ACADIAN ODYSSEY
By Jules M. Seletz



Acadians emigrated from France in the seventeenth century. During that century, these Acadians established a country in northeastern North America known as Acadia, living there for over one hundred years, until the French and Indian War in the middle of the eighteenth century. Upon the expulsion from their country by the English, Acadia became Nova Scotia, Cape Breton and Prince Edward Islands, and part of New Brunswick, Canada. Over the next fifty years, Acadians were scattered from Quebec, to the Upper Saint John Valley of Maine, along the eastern coast of the original colonies, back to France, and as far south as Louisiana and the Caribbean.

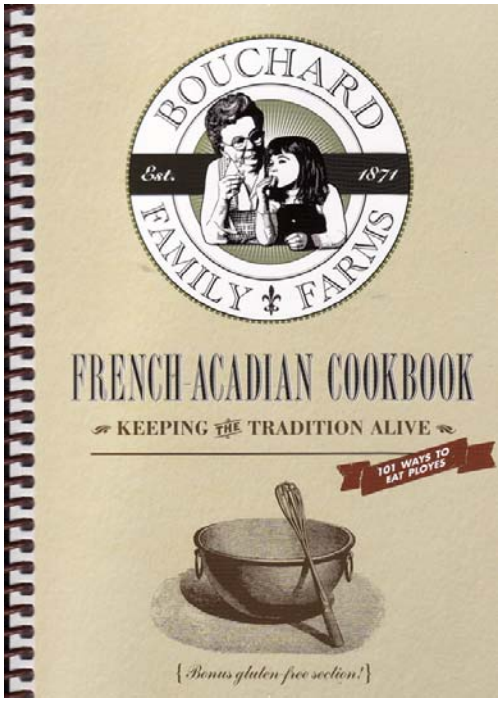
The book takes place from 1755 until 1955. It describes America's wars, the logging industry of the northeast United States, Maine's famous potato farms, the Roaring Twenties and the Great Depression of 1929. Eleven generations of family unity persevered through love of religion, language, and customs. They carried on with grit, determination, and ingenuity.

FRENCH-ACADIAN COOKBOOK: KEEPING THE TRADITION ALIVE

BOUCHARD FAMILY FARMS

Ployes! Pot en Pot! Tortierre! Creton! Raisin Pie! If you've ever had the good fortune to travel in Maine's St. John Valley, you're likely familiar with French-Acadian cuisine. Bouchard Family Farms, based in Fort Kent, Maine, has a new cookbook, ***French-Acadian Cookbook: Keeping the Tradition Alive*** that celebrates both the food and Maine's Acadian culture.

For generations the Bouchard Family has been milling a unique light buckwheat flour in order to prepare "Ployes" (rhymes with boys). A recipe based on the one created by the French Acadian exiles who settled in Northern Maine, Ployes are as elegant as a fine crepe, as hearty as a breakfast pancake and as versatile as any bread. Ployes are creating their own identity. From appetizers to main course to desserts and snacks, they keep finding new ways to make meals interesting. We invite you to try a product rich in taste and history. "C'est Magnifique!". Bouchard Family Farm Ployes Mix -- The first-ever original French Acadian buckwheat pancake mix produced in Northern Maine since 1983.



Visit Bouchard Family Farm's website for this and many other tasty products: http://www.ployes.com/

GUÉDRY & PETITPAS REUNION AGENDA

In less than 2 months we will hold our family reunion in Van Buren, Maine at the Acadian Village . Below is our final Agenda for the day. We published a great deal of information on the reunion, as well as an article on touring the Acadian regions of Maine and New Brunswick, in our Summer, 2013 and Spring, 2014 issues of GENERATIONS. Please visit our website below to find those issues. Just click on 'Newsletters' in the left-hand column and you will find them archived there.

http://freepages.genealogy.rootsweb.ancestry.com/~guedrylabinefamily

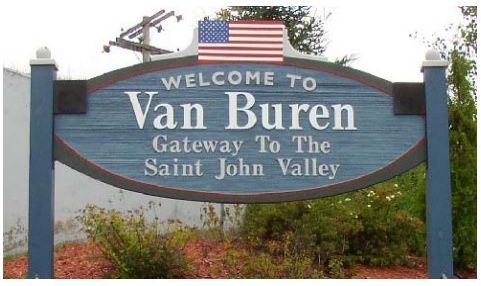
The Reunion registration form is attached to this newsletter for your convenience. It's not too late to make your plans to join us. We look forward to seeing familiar faces and meeting new family members.

GUÉDRY & PETITPAS REUNION
Saturday, 16 August 2014 – Van Buren, Maine

9:00 am	Registration & "Meet and Greet Cousins"
9:30 am	Opening
9:45 am	Chére Mom – an exciting Cajun band from South Louisiana featuring the Christine Guidry Law family
10:45 am	Break
11:00 am	French Mass at Notre Dame de l'Assomption Chapel (Acadian Village)
12:30 pm	Break
12:45 pm	Home-cooked Acadian meal (approximately $10 per person) [optional]
2:00 pm	Presentation - Madawaska Acadians: Their History and Heritage
2:45 pm	Break
3:00 pm	Induction of new honorees to the Circle of Distinction
3:30 pm	Break
3:45 pm	Presentation – Origins of the Guédry Family in France: Researching Our Roots in France
4:45 pm	Closing Ceremony
4:50 pm	Time to visit with cousins and enjoy displays

- Lots of time to meet and greet cousins during day
- Genealogical and historical displays to view
- Reunion souvenirs to purchase
- Family-related books to review and purchase

ALL TIMES ARE EASTERN DAYLIGHT TIME (EDT)



IN THE NEWS-HISTORICAL NEWS TIDBITS

Mrs. Antoine Guidry Succession
August 31, 1870
The Lafayette Advertiser

STATE OF LOUISIANA.
PARISH OF LAFAYETTE.
PARISH COURT.

WHEREAS Antoine Guidry of the parish of Lafayette has filed in said Court a petition, praying to be appointed Administrator of the Succession of Hortense Broussard, widow Antoine Sevenne Guidry, deceased.

Any person intending to make opposition to said appointment will file the same in said Court in the town of Vermilionville, within ten days from the publication of this notice, otherwise said appointment will be made.

Clerk's office Vermilionville, La., this 31st day of August A. D. 1870.

A. MONNIER,
Clerk of said Court.

Mareuse Guidry Case
August 26, 1922
Paper: Dallas Morning News, Dallas, TX

SIX ARRESTS MADE IN MAREUSE GUIDRY CASE.

Special to The News.

SHREVEPORT, La., Aug. 25.—Two more men, W. Duhon and Sam Plonski, have been arrested by Federal authorities on a charge of violating United States Judge Jack's injunction granted the Southern Pacific Railway, in connection with an attack upon Mareuse Guidry, an employe of the Southern Pacific at Lafayette, La., it was announced at the United States Marshal's office here Thursday.

Yesterday the Marshal announced the arrest of Leonce Billeaud, Ernest Domingue, Joseph Breaux and Ray Delahousaye, in connection with the attack upon Guidry, and their bonds today were set at $1,000 each and trial fixed for next Monday.

The Homer Guidry & Elizabeth Martin Family
August 8, 1973 **- Teche News, St. Martinville, LA**

GUIDRY HOUSEHOLD - This week's Picture from the Past recalls the family of Homer Guidry and Elizabeth Martin as they assembled on the porch of the family home in the Cecilia area back in 1893. From left are Lota Guidry, Andrew Robert, Martha Guidry, Eustiche Champagne, Jean Guidry, Andrian Robert, Mr. and Mrs. Homer Guidry, Mr. and Mrs. Noe Guidry and son (seated in chair) Jefferson, Arcade Daigle, Mr. and Mrs. Eulis Daigle, Toblan Daigle and Adiosca Daigle. Seated on the steps are Simon, Rose, Addelle, Leone and Emilian Guidry. The family dog was Lulu. Note the use of cut tree trunks for house supports and the v-shaped wooden gutters leading off to the right of the front roof. (Courtesy of Mrs. Abel Hanks)

IN THE NEWS-HISTORICAL NEWS TIDBITS

Charles Leo Labine Obituary
April 21, 1969
Omaha World Herald, Omaha, NE

Uranium Miner Labine Is Dead

Toronto (AP) — Charles Leo Labine, a partner with his brother Gilbert in discovering a uranium lode that supplied material for the first atomic bomb and other atomic devices, died Sunday. He was 81.

The Labine brothers made Canadian mining history and remained prominent in the industry for more than 40 years. Prospectors with no formal training, they discovered the original Eldorado pitchblende deposit — Canada's first — on Great Bear Lake in the Northwest Territories in 1931.

They raised cash to build the mine and managed it as the Eldorado Mining and Refining Co., until the Canadian government took it over in 1944.

Mr. Alex. Guidry
November 21, 1874
The Lafayetty Advertiser

Off for the Sea Coast.—A number of our fellow citizens left here last Thursday, on a pleasure excursion to the Vermilion Bay and the Gulf coast. Among the number are, Mr. F. Martin and family, Mr. H. Eastin and wife, Mr. Alex. Guidry and daughter, and several others. We wish them a pleasant and agreeable time, and that they may return home reinvigorated in health and spirits.

American State Papers
House of Rep. 14th Congress, Session One

No. 59.

674. David Guidry claims 480 superficial arpens of land, viz. twelve arpens front by forty deep, situated on the west side of Bayou Plaquemines Brulées, in the county of Opelousas; held under an order of survey to Pierre Guidry, dated the 4th March, 1788, and signed by Estevan Miro, then governor of Louisiana. The order of survey alone accompanies the notice. The claimant adducing no deed of sale from the original grantee, the confirmation is recommended to the legal representatives of Pierre Guidry.

No. 60

W. LaBean
June 13, 1916
Muskegon Chronicle (Muskegon, MI)

ST. IGNACE—A Michigan Central coach cleaner named W. LaBean, age 35, is held in Mackinaw City on a charge of murder as the result of the death of a fellow coach cleaner, Benjamin Burton, age 60. The two quarreled while at work two weeks ago and LaBean is alleged to have kicked Burton off a ladder and then to have pounded him so severely that his death resulted.

Les Guédry d'Asteur

What's in a name?

Guédry is the family to which you belong if your name is spelled Guédry, Guedry, Guidry, Gaidry, Guildry, Geddry, Grivois, Jeddry, Labine, LaBine, LaBean or any of several dozen variations. The original name of our family is believed to have been Guédry. We are all descendants of Claude Guédry & Marguerite Petitpas.

Here are some common and uncommon variant spellings of the name.

Guédry	Guiddry	Geddrie	Jeddrie	Labeen
Guedry	Guiddery	Geddry	Jeddry	Labene
Guedrie	Guiedri	Gedree	Jederie	Labine
Guedris	Guiedry	Gedrie	Jedrey	LaBine
Guidry	Guildry	Gedry	Jedrie	LaBean
Gudiry	Guildrie	Gettry	Jedry	LaBeau
Guidery	Guitry	Gidrie		Labeau
Guidrey	Gaidry	Gidry	Lledre	
Guidrie	Gaidrie	Grivois	Yedri	

Our **Petitpas** cousins likewise have several variations of their name including Petitpas, Pettipas, Petipas, Petitpa, Petit Pas and Pitts.

DUES REMINDER

Attached at the back of this issue is a membership application for renewing your membership in **Les Guédry d'Asteur**. Our dues are very reasonable at $6.00 for individuals and $10 for a family in 2014.

Please take a moment, complete the Membership Application, enclose a check and send it to the address on the application. It will help all of us do so much for the family. And, if you would like to join at one of the Benefactor Levels, it would allow us do even more.

Les Guédry d'Asteur is now on Facebook. Join us there and connect with other family members from all over the U.S. and CA. Feel free to post queries, photos, links, events or other items of interest to the family. Just search for 'Les Guédry d'Asteur' on Facebook to find our page.

Les Guédry d'Asteur

To share your ideas for the newsletter, contact:

Marty Guidry
6139 North Shore Drive
Baton Rouge, LA 70817
225-755-1915
guidryrm@cox.net

The Guédry-Labine Family Newsletter '***GENERATIONS***' serves as a focal point for family members to share and learn about us.

"***GENERATIONS***' newsletter is now in its 12th year. We hope to provide our readers with an interesting, informative and entertaining newsletter. Your input is always welcome and we look forward to another year of sharing family history and news with you.

Allie Guidry
txguidry2000@yahoo.com

Marty Guidry
guidryrm@cox.net

Les Guédry d'Asteur Officers and Committees

OFFICERS:
President - Martin Guidry (LA)
Vice-President - Elaine Clement (LA)
Secretary - Billy Harrell Guidry (LA)
Treasurer - Daniel "Chuck" Guidry (LA)

COMMITTEES:
Website - Becky Boggess (IA) - Chairperson
Annie Grignon-Labine (QU) - Translator
Elaine Clement (LA) - Translator
Martin Guidry (LA)

Genealogy - Daryl LaBine (FL/ON) - Chairperson
Bernard Geddry (AZ)
Mark Labine (MN)
Daniel "Chuck" Guidry (LA)
Martin Guidry (LA)

Finance - Cheryl Guidry Tyiska (MD) - Chairperson
Paul Labine (IL)
Marshall Woolner (OR)
Gloria Parrent (TX)
Chuck Guidry (LA)

Membership - Charlene Guidry Lacombe (LA) - Chairperson
Gayle Guidry (LA) - Special Projects
Warren Guidry (TX)

Sales - Cindy Guidry Herdt (WA) - Chairperson
Wayne Simoneaux (LA)
Billy Harrell Guidry (LA)

Publicity - Elaine Clement (LA) - Chairperson
Margaret Jeddry (MA)
Warren Guidry (TX)

Newsletter - Allie Guidry (VA) - Editor
Martin Guidry (LA)

CAFA Board Member - Jeanette Guidry Leger (LA)

Les Guidry d'Asteur
Membership Application
(Formulaire d'adhésion)

Name (Nom) _____
Last (Nom de famille) First (Prénom) Middle (Deuxième prénom)

Spouse (Épouse) _____
Maiden (Nom de jeune fille) First (Prénom) Middle (Deuxième prénom)

Children (Enfants) _____

Address (Adresse) _____
Street (Rue)

_____
City (Ville) State (État/Province) Zip Code (Code postal) (Pays)

Telephone (Téléphone) _____

Fax (Numéro de télécopieur) _____

E-mail Address (Courriel) _____

Hobbies or Special Talent _____
(Passe-temps ou talent particulier)

Type of Membership (Type de cotisation):

_____ Individual (Individuelle) $ 6.00 U.S. Dollars (Dollars américains)

_____ Family (Familiale) $10.00 U.S. Dollars (Dollars américains)

Benefactor Levels (Niveaux de bienfaiteur):

_____ dit Jovial Level $50.00 U.S. Dollars (Dollars américains)

_____ dit Labine Level $100.00 U. S. Dollars (Dollars américains)

_____ dit Grivois Level $500.00 U. S. Dollars (Dollars américains)

Please return form and payment to:
(Retournez le formulaire et le paiement à:)

Make check payable to: *Les Guidry d'Asteur, Inc.*
(Libellez le chèque à: *Les Guidry d'Asteur, Inc.)*

Les Guidry d'Asteur, Inc.
Charlene Guidry Lacombe
Membership Chair
226 Bulldog Lane
Iota, LA 70543

Les Guédry d'Asteur
REGISTRATION for 2014 REUNION
(L'enregistrement pour 2014 Réunion)
16 August 2014 – Van Buren, Maine

Name (Nom) _____
Last (Nom de famille) First (Prénom) Middle (Deuxième prénom)

Spouse (Épouse) _____
Maiden (Nom de jeune fille) First (Prénom) Middle (Deuxième prénom)

Children (Enfants) _____
First Names of Children (Prénoms de enfants)

Address (Adresse) _____
Street (Rue)

_____
City (Ville) State (État/Province) Zip Code (Code postal) (Pays)

Telephone (Téléphone) _____

E-mail Address (Courriel) _____

Number of People Attending (Le numéro de Gens qui assistent) _____

No. of Buffet Meals at Reunion (A l'intention de Manger des Repas à la Réunion) _____
(Buffet meal will cost $10-$12 per person / Le repas de buffet coûtera $10-$12 par la personne)
[This is not a commitment to purchase meals; we just need an estimate of potential meals needed]

Family - Parents and Children under 17 (Famille - Les parents et les Enfants sous 17):

_____ $40.00 Dollars (Canadian dollars for Canadian payments; U. S. dollars for U. S. payments) (Dollars canadiens pour les paiements Canadiens; Dollars américains pour les paiements américains)

Individual (Individuelle):

_____ $20.00 Dollars (Canadian dollars for Canadian payments; U. S. dollars for U. S. payments) (Dollars canadiens pour les paiements Canadiens; Dollars américains pour les paiements américains)

Please return form and payment to:
(Retournez le formulaire et le paiement à:)

Make check payable to: ***Les Guédry d'Asteur, Inc.***
(Libellez le chèque à: ***Les Guédry d'Asteur, Inc.)***

Les Guédry d'Asteur, Inc.
Martin Guidry, President
6139 North Shore Drive
Baton Rouge, LA 70817 USA