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Project Title: The clinical significance of trigeminal neuralgia nomenclature

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Summary (250 words max single spaced):

Introduction: A diagnosis of trigeminal neuralgia (TN) is often broadly applied to many neuralgic facial pains, while more stringent criteria are required for management decisions, outcome assessment, and pathophysiological correlations. Our aim was to evaluate existing classification systems of facial pain.

Methods: The study population was comprised of 534 Manitobans referred to neurosurgery for facial pain from 2001 to 2013. A retrospective chart review identified presenting features including pain distribution, nature, and duration. The recorded diagnoses (rDx) were then re-classified according to the International Classification of Headache Disorders (ICHD3) and Burchiel Classification System of TN1 and TN2.

Results: There was complete correlation between rDx and ICHD3 for diagnosis of typical TN (tTN) in 266(50%), atypical TN (aTN) in 37(7%) and atypical facial pain (AFP) in 59(11%). Another 135 (25%) had other facial pain diagnoses. The rDx of idiopathic trigeminal neuropathy (iTn) in 36(7%) was not classified in ICHD3. The application of Burchiel-TN1 criteria included patients with diagnoses including all those with tTN (266), but also cases of aTN (27), iTn (2) and IFP (8). Similarly Burchiel-TN2 included those with aTN (8), iTn (24), and IFP (15).

Conclusion: Stringent diagnostic criteria are vital for evaluation of all CFP conditions. We identified some omissions and inconsistencies among the existing classification systems relevant in discerning which patients may benefit from surgical treatments for TN.

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Introduction

The nomenclature of trigeminal neuralgia (TN) has taken many forms, from indiscriminate “facial pains” extrapolated from ancient writings [29], to t*ic douloureaux* in the 1750’s coined by Nicolas Andre [30] and Fothergill’s disease in 1770s.[29] The modern classification of TN is characterized as predominantly unilateral, recurrent, brief, lancinating electric shock-like pains which are confined to the trigeminal nerve distribution.[18]

The general characteristics of facial pain conditions have been defined and classified by the International Headache Society’s International Classification of Headache Disorders (ICHD). The diagnostic criteria for TN were originally quite broad, characterized by brief paroxysms of lancinating electric shock-like pain occurring within the trigeminal nerve distribution. More recently in the 3rd edition (ICHD-3), TN has been subcategorized to include: (i) purely paroxysmal classical trigeminal neuralgia (cTN-PP) and (ii) classical trigeminal neuralgia with concomitant persistent facial pain (cTN-CPFP) and these correspond to previously employed terminology of typical trigeminal neuralgia (tTN) and atypical trigeminal neuralgia (aTN).[18] Another widely utilized classification scheme was introduced by Burchiel, who also suggested two subtypes: (i) trigeminal neuralgia type-1 (TN1) defined by a predominance of episodic pain and (ii) trigeminal neuralgia type-2 (TN2) with predominance of persistent pain.[4, 10, 21]

Discrepancies between these two classification systems, ICHD-3 and Burchiel, are potentially significant. For example, when considering management options, particularly surgical interventions that may have utility limited to only some sub-types of TN, appropriate diagnosis is critical.[23] The analysis of treatment outcomes is also dependent on a discriminating and comprehensive classification system, such that results are reproducible and comparable. Ideally, a pain classification system will be based upon the underlying pathophysiology and distinguish between conditions that area potentially amenable to specific treatments such as surgery.

In the present study we reviewed a consecutive series of patients seeking neurosurgical consultation for facial pain and compared the commonly employed classification systems. Our aim was to assess the effectiveness of these systems in categorizing and distinguishing TN and other TN-like pain conditions

Methods

A consecutive series of all patients presenting with a primary compliant of facial pain for neurosurgical assessment at the Winnipeg Centre for Cranial Nerve Disorders (wCCND) from 2001 to 2013 were identified from a prospectively maintained clinical database. Only patients residing in Manitoba were included in this study. Each patient was categorized according to the recorded diagnosis (rDx) documented in the database.

While the diagnostic criteria and underlying pathophysiology are well established for tTN, there are some overlapping symptoms and potential for diagnostic confusion between aTN, idiopathic trigeminal neuropathy (iTn), and atypical facial pain (AFP). Among these patients a retrospective chart review was then conducted to extract clinical details relevant to general demographics and diagnostic features including pain characteristics, location, duration, intensity, as well as time to first neurosurgical consult.

Each patient was then re-categorized to a diagnosis defined by the criteria of the ICHD-3 and Burchiel classification systems, based upon the diagnostic features identified in chart review. Comparisons were then assessed between these diagnoses and the original rDx. The diagnostic criteria for each of these classification systems are shown in table 1.

Statistical analyses were conducted using IBM SPSS Statistics version 24 and Microsoft Office Excel 2013, including independent t-tests and two-tailed chi-squared (χ^2) used for comparing

continuous and categorical variables, respectively. Differences were considered significant for p-values <0.05.

The study received approval from the University of Manitoba Research Ethics Board, ethics number HS16272 (H2013:145), and Undergraduate Medical Student Research Committee.

Results

There were a total of 534 Manitoba patients assessed for the primary complaint of facial pain over the 13 year study, a rate of 3.4/100,000/year. The most common rDx was tTN in 50%, followed by AFP, aTN and iTn, 11%, 7%, and 7%, respectively. Another 25% of patients had other diagnoses such as unspecified headache (6%), MS-related TN (6%), post-traumatic TN (4%) and a variety of other conditions (<2% each) shown in table 2. Chart review was completed for 35 of the 37 aTN, 40 of the 59 AFP, and 26 of the 36 iTn, while charts were not available for the remaining patients. Two patients, 1 from iTn and AFP were excluded from analysis as they were subsequently diagnosed with multiple sclerosis as the underlying cause of facial pain.

Demographics

The demographic data for aTN, AFP, and iTn are presented in table 3. There was an appreciable predominance of female patients afflicted by all three disorders, with no significant difference between the diagnoses ($\chi^2 = 0.774$, $p = 0.679$).

The mean ages of symptom onset from youngest to oldest were AFP at 43 ±12.4 years (range 14-63), then iTN at 46.5 ±14 (range 21-76), then aTN at 56.7 ±14.4 (range 33-88), figure 1. There were significant differences between aTN versus AFP and aTN (p= 0.000 and 0.007, respectively). The age difference between AFP and iTN was not significant (p-value = 0.364).

The average duration, in months, of pain from onset to first neurosurgical consult was shortest for aTN (56.8 ±48.6, range 5-186) then AFP (69 ±64.7, range 6-251) then iTN (127 ±197.7, range 2-752). There was a statistically significant difference between duration of the iTn and aTN only (p=0.048), while the differences in duration between AFP versus iTn and aTN were not (p=0.095 and 0.470, respectively).

Pain Characteristics

Atypical Trigeminal Neuralgia: Episodic pains less than 2 minutes were described by 100% of the cohort at initial consult, together with some level of daily, longer lasting pain. The pain severity was reported as severe or 7-10 range on the visual analog scale. Pain was right sided in 23 of 35 (66%), and bilateral but non simultaneous in one patient. The pain was confined to the trigeminal distribution and most commonly confined to V_3 (37%), but in others presented in various trigeminal divisions; V_{2+3} in 9 (26%), V_{1+2} in 6 (17%), V_2 in 4 (11%), V_{1+2+3} in 2 (6%), and V_1 in 1 (3%) patient, table 4.

Atypical Facial Pain: All 40 (100%) described daily and long-lasting pain (>2 hours) of varying severity. Predominantly severe pain was noted in 30 (75%), 7 (17%) described moderate and 3 (8%) only mild pain symptoms. Additionally, less prominent episodic, brief pain attacks were also described by 25 (63%). Pain laterality was bilateral in 17 (43%), left sided in 14 (35%) and right sided in 9 (23%). The distribution of pain extended beyond the trigeminal nerve territory and was poorly localized in 28 (70%) as shown in table 4. Among those 12 (30%) patients presented with pain limited to the trigeminal distribution, the primary pain was not neuropathic in nature. No patients had somatization disorders, although 15 (38%) reported anxiety and/or depression difficulties.

Idiopathic Trigeminal Neuropathy: These patients had a persistent neuropathic symptoms consisting of predominantly subjective numbness, deep, aching, or burning pain, as well as less predominant brief paroxysmal pains, all confined to the trigeminal distribution. Most, 24 (92%), reported their most significant pain as constant or long lasting rather than brief or paroxysmal. While the majority of patients reported severe symptoms (58%), 7 (27%) experienced moderate and 4 (15%) had mild symptoms. Pain was left sided in 11 (42%), right sided in 9 (35%) and bilateral in 6 (23%). No patients reported solely V_1 symptoms, however; 9 (35%) had V_2, 6 (23%) had V_{2+3}, 3 (12%) had V_3, 2 (8%) had V_{1+2}, 2 (8%) had V_{1+2+3} involvement, and 4 (15%) had symptoms poorly localized within the trigeminal distribution, table 4. Neurological examination demonstrated trigeminal sensory abnormalities in 15 (58%) and no underlying etiology was evident (table 5).

Classification of Pain Conditions

The application of the ICHD-3 criteria to each patient matched their rDx for tTN, aTN, and AFP with CTN-PP, CTN-CPFP, and PIFP, respectively. Other rDx had corresponding conditions within the ICHD-3, such as tumor related pain, post traumatic pain, etc, with the exception of the rDx of iTn for which no corresponding diagnosis was available in the ICHD-3.

When the Burchiel classification system was applied, patients with rDx of tTN (and CTN-PP) all met the diagnostic criteria for TN1, although not exclusively (table 6). Patients with aTN (and CTN-CPFP) met criteria for TN1 in 77% and TN2 in 23% while those with iTn met criteria for TN1 in 8% and TN2 in 92%. While the Burchiel system identifies a category of “AFP”, it is assigned only to:

> those patients with unequivocal evidence of a somatoform pain disorder that can be objectively diagnosed by psychological testing...[limited to patients] with simultaneous facial pain, pain spreading well outside the trigeminal distribution, multiple pain complaints in other body regions, and diagnostic clustering with conditions such as fibromyalgia and chronic fatigue syndrome.[4]

No patients in this series met these criteria of Burcheil-AFP classification, while those with an rDx of AFP met Burcheil criteria for TN1 in 20% and TN2 in 38%. There were 17 (42%) with an rDx of AFP that did not meet criteria for any Burchiel classification diagnoses, as their facial pain symptoms extended beyond a trigeminal distribution and therefore not a form of TN but also not fulfilling the Burcheil-AFP criteria (table 6).

Discussion

The present study allowed us to examine the nature of facial pain conditions among Manitoba residents referred for neurosurgical evaluation and treatment. The wCCND is the sole provider of such services in the province, such that a complete representation of these patients was available for analysis. Not surprisingly, TN was the most common condition diagnosed, representing 50% of referred patients. These patients have excellent potential to benefit from a variety of neurosurgical procedures effective specifically for TN.[15] One significant finding was that a large proportion of patients referred to the wCCND for facial pain had diagnoses that were not amenable to surgical interventions. This finding highlights the importance of accurate diagnoses, such that only patients with appropriate conditions are offered surgeries while others are redirected to non-surgical treatment options.

Accurate diagnosis of TN and facial pains has been recognized as a challenge.[22] The condition is relatively rare, with an incidence of trigeminal neuralgia 4.3/100,000/year in Rochester, Minnesota.[20] That study was based upon a comprehensive medical records

linkage-system in the Mayo Clinic, where diagnoses were determined by neurological specialists. More recent epidemiological data reported dramatically higher incident rates, ranging from 8 to 29.5/100,000/year.[9, 11, 13, 14, 22, 26] These reports were based upon electronic medical record reviews or survey information, each associated which varying or no levels of specialist validation. It is evident that less discerning assessment or classification of facial pain diagnoses may lead to over diagnosis of TN.

There were 25% of patients in our series with facial pain conditions that had TN-like conditions, including aTN, iTn, and AFP. Additionally, another 25% had other defined conditions readily discernible from TN and could be considered for diagnosis-specific interventions. Among the TN-like conditions, only aTN has a similar underlying pathophysiology of neurovascular compression upon the trigeminal nerve root.[8, 24] Both tTN and aTN are potentially amenable to microvascular decompression surgery (MVD), although results for aTN are not generally as favourable as for tTN.[32] The alternative surgical option of rhizotomy procedures, useful for tTN, is considered less effective for aTN where the pre-existing persistent pain component may be aggravated by the induced nerve injury. Overall, any degree of pre-existing persistent or constant pain has been found to be a negative predictor of surgical outcome.[19, 25, 28, 32, 34] The symptoms of iTn are similar to those caused by pathologies that damage the trigeminal nerve, such as tumors, herpetic zoster infections and trauma. As in those conditions, rhizotomy procedures to intentionally injure the nerve are contraindicated as these potentially will aggravate the existing neuropathic-like pain. It is therefore evident that patterns with iTn should not be misclassified under a TN diagnosis and undergo misdirected TN-specific interventions. Treatment of iTn instead relies on medical therapy akin to that used for neuralgia/neuropathic pain disorders, such as anticonvulsants and tricyclic antidepressants.[6, 13, 14, 27, 33]

For patients with AFP, surgery is contraindicated. An evidence-based medicine review suggesting little to no benefit from surgical interventions.[7] Similarly the Canadian Agency for Drugs and Technologies in Heath in their review identified that surgical options were non-beneficial, with more high-powered studies being required.[5] Therefore, it is crucial for neurosurgeons to recognize those with AFP and avoid surgery for these patients that provides no potential benefit and may be harmful.

Our study identified and described clinical features of patients with iTn diagnosed in our centre. This conditions has unique features discernable from other facial pain disorders, although is not described in the ICHD-3 system. The closest approximation of iTn in the ICHD-3 was “*Painful trigeminal neuropathy attributed to other disorder”* and differs in that iTn has no discernible underlying condition or cause. There are have been few reports of iTn-like conditions found in literature. For instance, Hughes reported in 1958 three patients, two women and one man with pain and numbness throughout the trigeminal distribution progressing over years since initial onset.[17] Surgical intervention was undertaken in all three patients to control pain via division of the trigeminal sensory root, when it was discovered that sensory root attrition and thickened arachnoid was present. The surgeries resulted in complete relief of pain and paresthesia in two patients and one case resulted in persistent burning pain. Another case report outlined the peripheral trigeminal sensory disturbances, including tingling and discomfort, of the buccal and lingual nerve believed to be due to muscle compression.[1] Treatment included oral splinting and physiotherapy to reduce dental attrition which provided marked improvement. Similarly Shotts et al identified two case reports of an idiopathic trigeminal sensory neuropathy (ITSN).[31] ITSN was later classified into two forms, acute and chronic by Flint in 1990.[12] Based on these reports the chronic form was found to have associations with connective tissue disorders while the acute form had a much more favourable prognosis with postulated associations to infections. Our own series of 26 patients allowed for the comparison to other TN-like conditions and establishment of recommendations for a diagnostic criteria for iTn.

The comparison of rDx assigned at the wCCND with the ICHD and Burchiel classification system diagnoses demonstrated some important differences. Overall we found our rDx matched the ICHD diagnoses, except for iTn which is not described in the latter. The Burchiel system, however, grouped patients with differing diagnoses into either TN1 or TN2 categories, and also failed to categorize some with AFP. Burchiel introduced the TN1 definition to encompass patients with tTN and aTN, suggesting the former generally evolved to the latter and should be considered a single entitiy.[3, 4, 10, 28] In our series, patients with aTN were distinct from those with longstanding tTN that never developed constant pain. While the Burchiel-TN1 category simplifies diagnostic criteria, it fails to differentiate between actual pain conditions that have different response rates to interventions and potentially have differing underlying pathophysiology. At the extreme, the Burchiel classification system may lead to misdirected surgeries. For example, in our review we found 8% of iTn and 20% with AFP patients met criteria for Burcheil-TN1. Neither of those conditions are amenable to surgical interventions useful for tTN and aTN. In noting this, we concur with prior suggestions that the Burchiel classification system is insufficiently discerning of disorders when considering surgical treatments.[2]

Conclusion

There are some clinically significant applications of the findings from our study. Firstly, we can emphasize the importance of accurate diagnosis for TN as many patients referred to neurosurgery with “trigeminal neuralgia” were found to have different diagnoses and different treatment approaches were indicated. Secondly, we identified and defined a condition of iTn, not previously categorized. This condition has clinical similarities to other trigeminal nerve damaging conditions, therefore management should rely on non-invasive medical therapy. Finally, we have demonstrated that the recently popularized Burchiel classification system for TN may lead to erroneous diagnoses for non-TN conditions such as AFP and iTn, both of which should not managed through the same surgical interventions as tTN and aTN.

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Table 1. Diagnostic criteria utilized at Winnipeg Centre for Cranial Nerve Disorders (wCCND) and according to the International Classification of Headache Disorders (ICHD-3), Burchiel Classification.

wCCND	
tTN	Brief, lancinating, electric shock-like paroxysms, confined to the trigeminal nerve distribution.
aTN	Brief, lancinating, electric shock-like paroxysms superimposed over persistent deep, aching, throbbing pain, confined to the trigeminal nerve

	distribution.					
iTn	Neuropathic pain of an aching, burning, tingling, and/or numb nature limited to the trigeminal nerve distribution, with presence of typical trigeminal paroxysms occurring at some point in disease onset, and notable disease evolution. Symptoms not accounted for by a diagnosis of aTN or AFP and without underlying cause (eg. Trauma, ms, tumor, etc)					
AFP	Persistent pain, of a deep, aching, nagging, throbbing nature not necessarily confined to a specific cranial nerve distribution, associated with psychiatric conditions, and/or are affected by stress or emotional state. Episodic pains may be present.					
ICHD-3						
Classical trigeminal neuralgia (CTN)	A. At least three attacks of unilateral facial pain fulfilling criteria B and C	B. Occurring in one or more divisions of the trigeminal nerve, with no radiation beyond the trigeminal distribution	C. Pain has at least three of the following four characteristics • Recurring paroxysmal attacks lasting from a fraction of a second to 2 minutes. • Severe intensity • Electric shock-like, shooting, stabbing or sharp in quality • Precipitated by innocuous stimuli to the affected side of the face	D. No clinically evident neurological deficit	E. Not better accounted for by another ICHD-3 diagnosis	
CTN-purely paroxysmal (CTN-PP)	A. Recurrent attacks of unilateral facial pain fulfilling criteria for Classical trigeminal neuralgia	B. No persistent facial pain between attacks	C. Not better accounted for by another ICHD-3 diagnosis			
CTN-with concomitant persistent facial pain (CTN-CPFP)	A. Recurrent attacks of unilateral facial pain fulfilling criteria for Classical trigeminal neuralgia	B. Persistent facial pain of moderate intensity in the affected area	C. Not better accounted for by another ICHD-3 diagnosis			
Persistent idiopathic facial pain (PIFP)	A. Facial and/or oral pain fulfilling criteria B and C	B. Recurring daily for >2 hours per day for >3 months	C. Pain has both of the following characteristics: • Poorly localized, and not following the distribution of a peripheral nerve • Dull, aching, or nagging quality	D. Clinical neurological exam is normal	E. A dental cause has been excluded by appropriate investigation	F. Not better accounted for by another ICHD-3 diagnosis
Burchiel						
TN1	Sharp, shooting, electrical shock-like, episodic pain >50% of the time					
TN2	Aching, throbbing, burning, constant pain >50% of the time					
AFP	Patients with unequivocal evidence of somatoform disorder that can be objectively diagnosed by psychological testing					

Table 2. Diagnosis of patients referred to neurosurgery with a primary complaint of facial pain. (tTN = typical trigeminal neuralgia, AFP = atypical facial pain, aTN = atypical trigeminal neuralgia, iTn = idiopathic trigeminal neuropathy, other included diagnoses of tumor-related TN, multiple sclerosis-related TN, post-traumatic TN, etc.)

Recorded Diagnosis	Number of Patients	%

tTN	266	50
AFP	59	11
aTN	37	7
iTn	36	7
Other	147	25
Total	534	100

Table 3. Patient demographics for aTN, AFP, and iTn diagnosed patients (aTN = atypical trigeminal neuralgia, AFP = atypical facial pain, iTn = idiopathic trigeminal neuropathy).

	aTN	**AFP**	**iTn**
Sex			
M	11 (31%)	10 (25%)	9 (35%)
F	24 (69%)	30 (75%)	17 (65%)
Total	35	40	26
Age at onset			
Mean (SD)	56.7 (±14.4)	43.5 (±12.4)	46.5 (±14)
Range	33-88	14-63	21-76
Duration to 1st NS consult (mo.)			
Mean (SD)	56.8 (±48.6)	69 (±64.7)	127 (±197.7)
Range	5-186	6-251	2-752

Table 4. Pain characteristics as described during neurosurgical consult (aTN = atypical trigeminal neuralgia, AFP = atypical facial pain, iTn = idiopathic trigeminal neuropathy).

	aTN	**AFP**	**iTn**
Pain			

Episodic	35 (100%)	25 (63%)	16 (62%)
Constant	35 (100%)	40 (100%)	24 (92%)
Episodic and constant	35 (100%)	25 (63%)	16 (62%)
Severity			
Mild (0-3/10)	0 (0%)	3 (8%)	4 (15%)
Moderate (4-6/10)	0 (0%)	7 (17%)	7 (27%)
Severe (7-10/10)	35 (100%)	30 (75%)	15 (58%)
Symptom location			
Left	11 (31%)	14 (35%)	11 (42%)
Right	23 (66%)	9 (23%)	9 (35%)
Bilateral	1 (3%)	17 (43%)	6 (23%)
V_1	1 (3%)	0	0
V_2	4 (11%)	3 (7%)	9 (35%)
V_3	13 (37%)	1 (3%)	3 (12%)
V_{1+2}	6 (17%)	0	2 (8%)
V_{2+3}	9 (26%)	5 (13%)	6 (23%)
V_{1+2+3}	2 (6%)	3 (7%)	2 (8%)
Poor loc.	0	28 (70%)	4 (15%)

Table 5. Idiopathic trigeminal neuropathy symptoms according to patient reports at neurosurgical consult.

	Number of patients
Persistent	24 (92%)
Dull	5 (19%)
Burning	5 (19%)
Tingle	7 (27%)
Numb	14 (54%)
Ache	9 (35%)
Episodic	16 (62%)
Needle	1 (4%)
Throb	3 (12%)
Lancinating (stabbing, jabbing, shooting)	10 (38%)
Electric	4 (15%)
Sharp	5 (19%)
Neurological exam	
Abnormalities	13 (50%)
Disease progression	
Change in region	15 (58%)
Episodic to include persistent	10 (38%)
Persistent to include episodic	3 (12%)
Progression to include numbness	6 (23%)

Table 6. Comparison of recorded diagnosis with ICHD and Burchiel classification systems.

	tTN	**aTN**	**AFP**	**iTn**

TN1	266 (100%)	27 (77%)	8 (20%)	2 (8%)
TN2	0	8 (23%)	15 (38%)	24 (92%)
Undefined	0	0	17 (42%)	0
Total	266	35	40	26

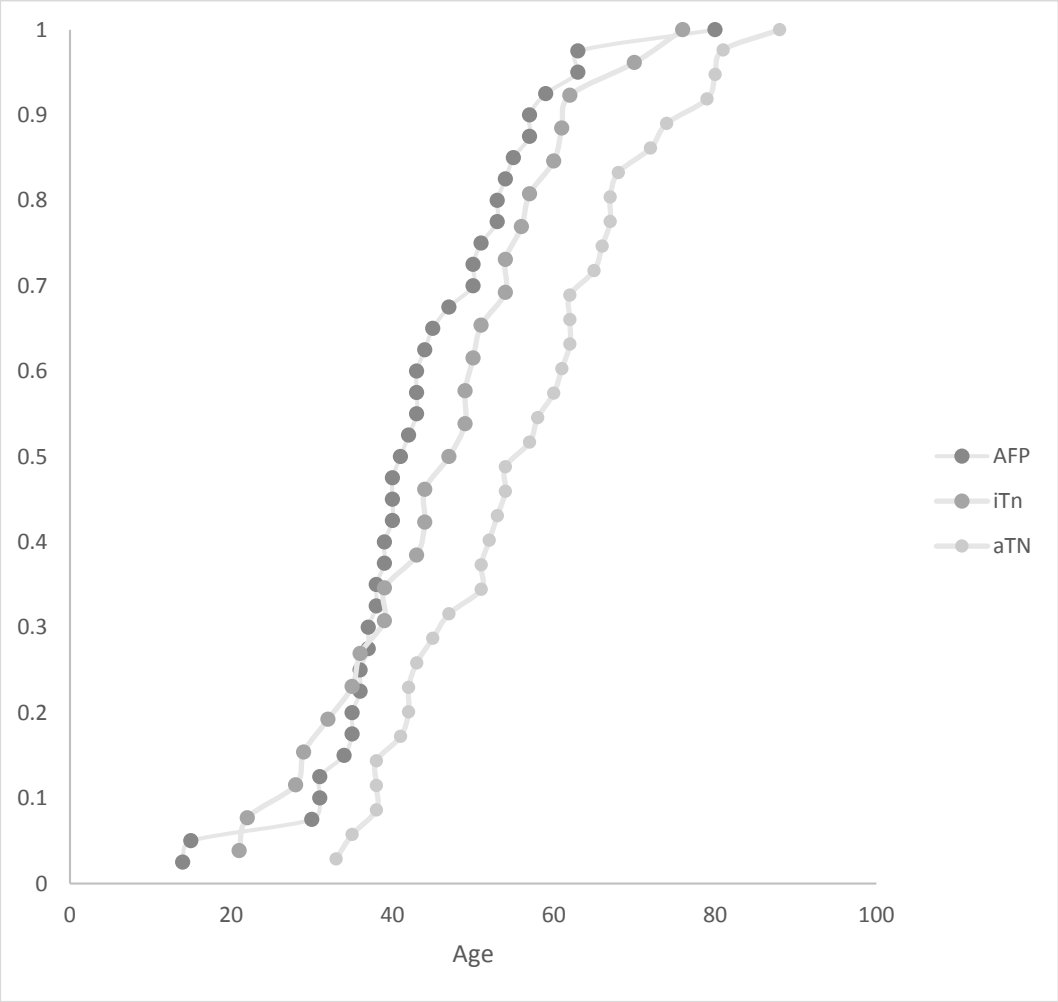

Figure 1. The cumulative distribution of age of onset of symptoms for AFP(dark grey), iTn (grey), and aTN(light grey).