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The history of Jervell and Lange-Nielsen syndrome

MEDICAL HISTORY

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Background

First-time documentation of rare diseases is normally in the form of case reports. These are typically based on unexpected observations by vigilant clinicians and lead to further research on prevalence and aetiology. One of the best-known Norwegian examples is Jervell and Lange-Nielsen syndrome.

Material and method

We conducted a literature review using systematic database searches and historical analysis.

Results and interpretation

The Norwegian internal medicine specialists Anton Jervell (1901–87) and Fred Lange-Nielsen (1919–89) published an article in 1957 in the *American Heart Journal* describing a Norwegian family in which four of the six children were deaf and experienced recurrent syncope. Three of the children died suddenly. This rare condition was the first evidence of prolonged QT intervals on an ECG as a risk factor for sudden cardiac death.

From the 1960s, the condition became commonly referred to as Jervell and Lange-Nielsen syndrome. Jervell was the leading expert in describing the syndrome, while Lange-Nielsen subsequently specialised as a pulmonologist. Johan Kloster (1901–77), a senior consultant at Aust-Agder Central Hospital in the Norwegian town of Arendal made a significant contribution to the work but was not credited as a co-author on the article.

The discovery of Jervell and Lange-Nielsen syndrome has had a profound impact on research into heart rhythm disorders and genetics. The genetic aetiology of the syndrome was identified in the 1990s, and several causes of prolonged QT intervals have since been uncovered. Jervell and Lange-Nielsen syndrome is now considered a subgroup of long QT syndrome.

Jervell and Lange-Nielsen syndrome is one of the best-known medical syndromes named after Norwegian doctors (1). It is a life-threatening hereditary condition characterised by deafness and prolonged QT intervals (2). The QT interval is measured on an electrocardiogram (ECG) from the start of the QRS complex to the end of the T wave and reflects the duration of the heart's contraction phase (systole). The more prolonged the QT interval, the greater the risk of arrhythmia in the form of ventricular tachycardia, which can cause fainting and, in the worst cases, cardiac death. Despite the significance of the discovery, little has been written about the history of this syndrome. Who were the doctors who first documented it, what are its characteristics, how was it discovered and what were the consequences of the discovery?

Material and method

We searched PubMed, Google Scholar, Web of Science, the Nordic media archive Retriever, the digital archives of the *Aftenposten* newspaper and the National Library of Norway's online database. The syndrome was entered in MeSH (Medical Subject Headings) under its own indexing term in 2001 (3). We also searched the University of Oslo's Medical Faculty archive at the National Archives of Norway for material associated with Jervell's doctoral thesis defence in 1936 and the announcement of the professorship in internal medicine in 1955 (4).

The original patient records have not been found, either at the hospitals in Tønsberg (Jan Erik Otterstad, personal communication) or Arendal (Torstein Gundersen, personal communication). They have also not been deposited with the Norwegian Health Archives in Tynset (Kari Nytrøen, personal communication) and are therefore probably lost.

The significance of Jervell and Lange-Nielsen's discovery was analysed using an inductive approach to the relationship between their published article and the subsequent knowledge advancement on prolonged QT intervals (5).

The doctors

Anton Jervell (1901–87) (Figure 1) grew up in Kristiania (now called Oslo), qualified as a medical doctor in 1925 and as a specialist in internal medicine in 1933, and was awarded his doctoral degree in 1936 (6–8). His doctoral thesis, written in German, was based on research conducted at Department 8 of Ullevål Hospital in Norway in the period 1931–1935. The focus of the thesis was electrocardiographic findings in myocardial infarction (9). It was described as pioneering work, as this was a new and little-known disease. Several weeks could pass between the admission of patients with myocardial infarction (10), and ECG had only been introduced as a diagnostic method a few years earlier (11).

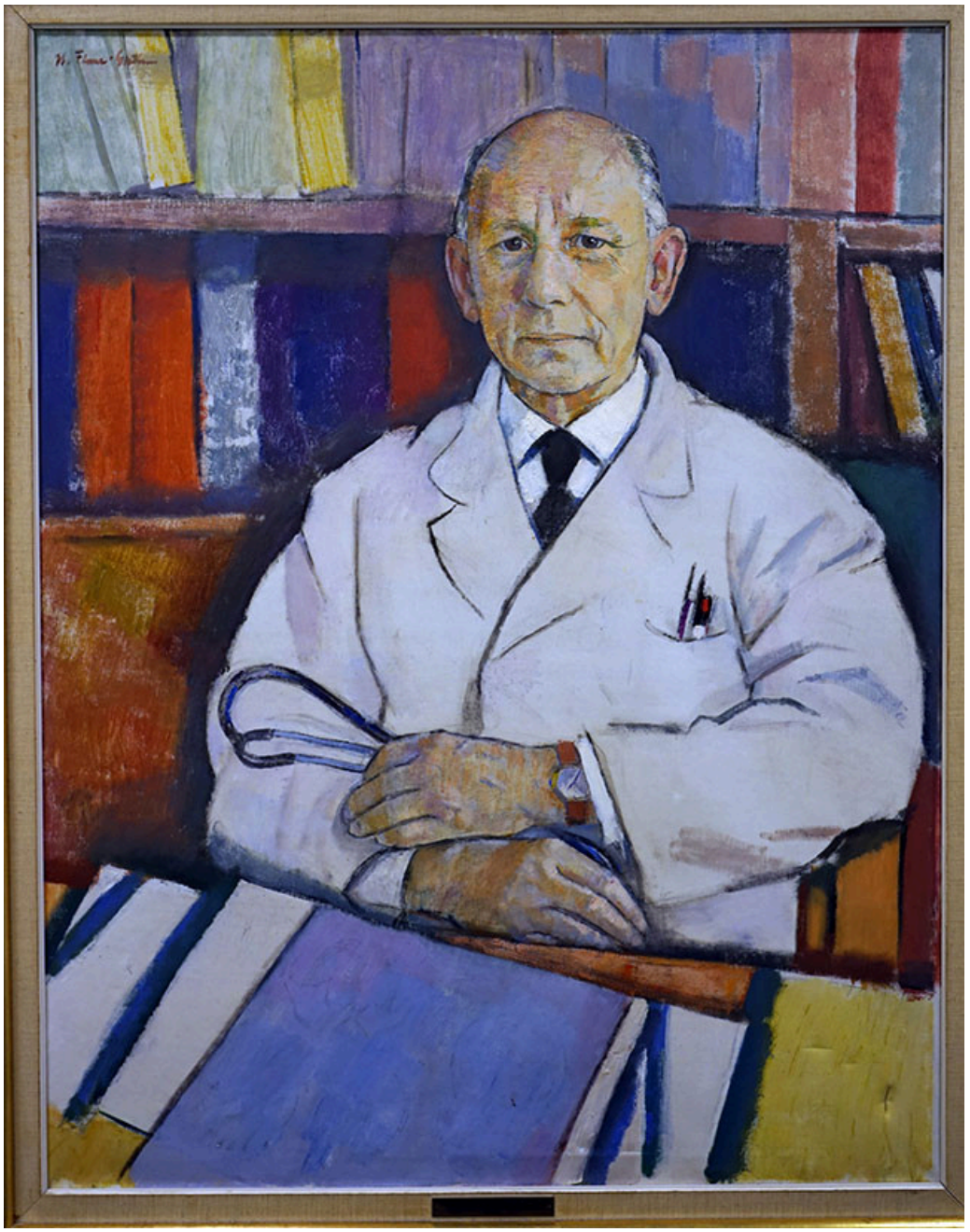

Figure 1 Anton Jervell (1901–87), painted by Hans Finne-Grønn (1903–2001) in 1967. The painting is on display in Building 3 (the medical building), Ullevål Hospital. Photo: Svein Skog, Oslo University Hospital.

Jervell was a locum doctor at Bærum Hospital, Ullevål Hospital and Rikshospitalet in Norway, and had study exchanges in locations like Vienna, London, Paris and the United States. In 1938, he was appointed as senior consultant at the newly established medical department at the hospital in Tønsberg (Figure 2) (12).

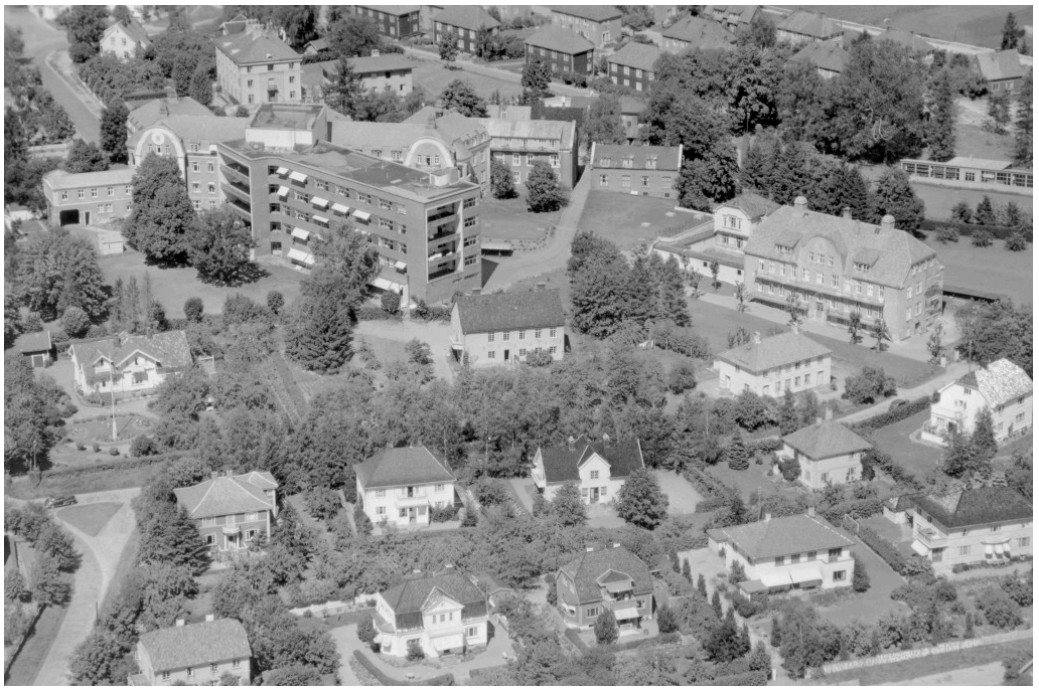

Figure 2 *Vestfold County Hospital in 1950. Jervell lived in the white house farthest right on the photo. The brick building in the centre of the image was built in 1938, with the surgical department on the lower two floors and the new medical department on the upper two floors. The photo is from the Medical History Society (*Medisinsk historielag*), Vestfold Hospital.*

Jervell was involved in a wide range of activities in the county of Vestfold. He was chairman of the Tønsberg Red Cross and tried to help the Jews in the Berg concentration camp. He was arrested by the Germans in September 1943 but was released about two months later (13. 357). After the war, he became involved in local politics as a Labour Party representative in Tønsberg City Council and was appointed director of the hospital in 1947. In 1941, Jervell was the chief editor of a textbook for nurses, which dominated nursing education for nearly 30 years.

In 1955, two professorships in medicine were announced at Departments 7 and 8 at Ullevål Hospital. The professorships were combined with senior consultant positions, and Jervell was one of seven applicants. The Faculty of Medicine at the University of Oslo established a scientific committee consisting of five professors: one from Helsinki, Copenhagen and Malmö, and two from Norway. The expert evaluations filled 220 pages of the university's annual report (14).

The reviewers pointed out that Jervell's published works from the last 15 years were 'nowhere near' the level of his doctoral thesis (14, p. 395). His scientific career had not shown the 'desired growth and development, quite the opposite'. However, they understood why this had been the case. During the war, he had not published anything, and afterward, his capacity had been 'substantially taken up with efforts to establish a good department at a good central hospital, where he had made an outstanding contribution'. In the administrative field, his efforts were considered 'groundbreaking' (14, p. 535), and he had also demonstrated quite unique qualifications as a university lecturer (14, p. 570).

The dean believed that Jervell was one of the top two candidates, but the 'overall impression from the statements' was so even that it was only natural for the faculty to 'place them on an equal footing' (4). The executive committee of

Oslo City Council therefore had the final say. Jervell was appointed as a senior consultant in Department 8 and took up his position in January 1957, succeeding Carl Müller (1886–1983).

Fredrik (Fred) Lange-Nielsen (1919–89) (Figure 3) also grew up in Norway's capital city. He was the eldest of three siblings, all of whom had artistic talents. He became a skilled jazz musician and was a member of several orchestras (15, 16). He was a recording studio musician, gave radio lectures and appeared in TV programmes.

Figure 3 Fred Lange-Nielsen (1919–89) painted by Ville Aarseth (1899–1985) in 1979. The painting is on display in Building 3 (the medical building), Ullevål Hospital. Photo: Svein Skog, Oslo University Hospital.

In 1937, Lange-Nielsen began studying medicine. He was arrested in October 1944 and detained for nearly six months (13, p. 428). He qualified as a medical doctor in 1947, a specialist in internal medicine in 1956 and a pulmonologist in 1964. In the early years of his medical career, he undertook short-term work in private practice in Gjøvik and at Lovisenberg, Haukeland, Gaustad and Rikshospitalet hospitals.

From 1953 to 1956, he worked at the hospital in Tønsberg as a junior registrar and locum. The following year, he began working at Ullevål Hospital, where he remained as a pulmonologist for nearly his entire career. He became particularly involved in allergology, and it was through his initiative that the Laboratory for Clinical Allergology was established in Department 9 in the 1960s (17). This was the only hospital outpatient facility for adults with allergic respiratory diseases and asthma in Norway for many years (18). He was one of the founders of the Norwegian Asthma and Allergy Association in 1960 and served as head of the medical council there for many years. In the 1970s, he spearheaded the establishment of an interdisciplinary medical school for asthma.

Fred Lange-Nielsen had a broad range of interests and was involved in many different initiatives. In 1955, he advocated for Norway to lead a campaign against boxing. He believed the consequences of minor head injuries were significantly underestimated (19). He was active in the fight against apartheid, and in 1963, he initiated and was the first chairman of Norway's Crisis Fund for South Africa (*Krisefondet for Sør-Afrika*). The following year, the International Defence and Aid Fund for Southern Africa (IDAF) was established, and he was elected vice president (20).

Jervell and Lange-Nielsen syndrome

In October 1953, a nine-year-old boy was admitted to the medical department at Vestfold Central Hospital due to repeated fainting episodes. The boy was born deaf, and since the age of three, he had experienced several fainting episodes of up to 2–3 minutes duration. The episodes often occurred after physical exertion. During the episodes, he was sometimes pale and other times blue in the face, but he did not have seizures. Apart from these episodes, he was healthy and in normal physical condition, except for occasional complaints of palpitations and chest pain (1, 21).

Because of these episodes, he was examined by Johan Kloster (1901–77), a senior consultant at Aust-Agder Central Hospital in the Norwegian municipality of Arendal, in July 1953. The only abnormal finding was a prolonged QT interval on the ECG. The episodes could suggest epilepsy, but trial anti-epileptic treatment had no effect (1, 21).

During his four-week hospital stay in Tønsberg, no abnormalities were found in the clinical examination, heart X-ray or various laboratory tests. The same change in the ECG was observed (Figure 4). No treatment was initiated (1, 21).

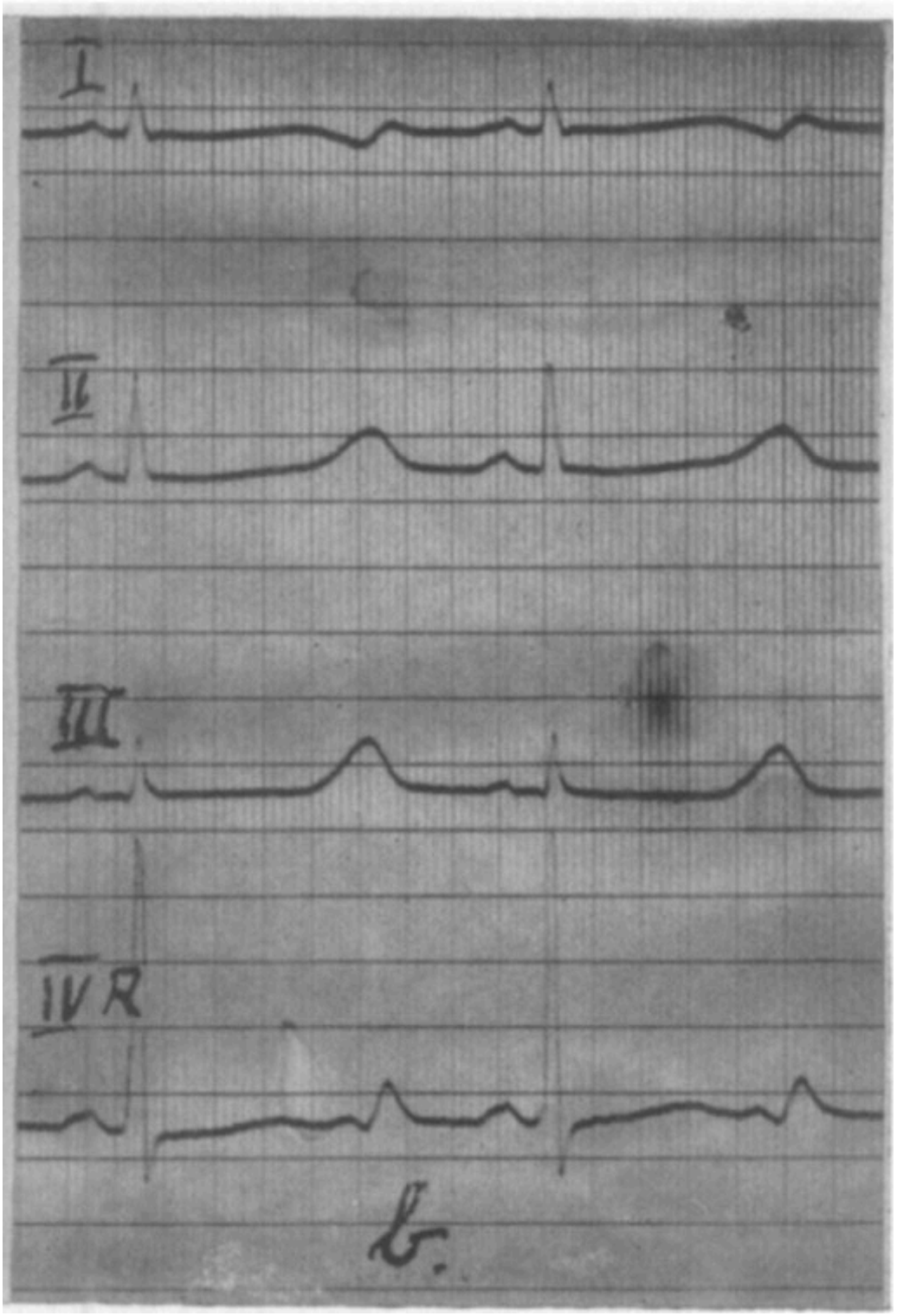

Figure 4 *Facsimile from the article in the* American Heart Journal *in 1957 (1). The T wave (above the 'b' in the image) is significantly delayed: it should normally occur well before the midpoint between the sharp QRS complexes.*

Just one week after his discharge in November 1953, the boy had another fainting episode, and this time he did not regain consciousness. By the time he arrived at the hospital, he was dead. The autopsy revealed no explanation for his condition, particularly with regard to his heart (1, 21).

The boy's mother had given birth to six children over a period of ten years. Four were deaf and had experienced fainting episodes that began at ages 3–5. Two girls died at the age of five, in September 1953 and February 1956 (21). Three

children had prolonged QT intervals, while the fourth had not been examined before she died. Two siblings and the parents had normal hearing, normal ECG findings and no fainting episodes.

In November 1956, Jervell and Lange-Nielsen submitted a manuscript describing the case of the four children to the *American Heart Journal*, which was published in July 1957 (1). The authors concluded that the children must have had the same previously unknown heart disease. The combination of congenital deafness and a heart condition with distinct ECG changes and sudden death in multiple siblings could not have been a mere coincidence; however, the doctors had no explanation for the link: 'The conformity of the clinical pictures and of the electrocardiographic abnormalities makes it evident that the 4 deaf-mute children were all suffering from the same kind of heart disease. The nature of this, however, is quite obscure' (1, p. 66).

Jervell and Lange-Nielsen concluded the article with: 'The unusual clinical symptoms, the exceptional electrocardiographic findings, and the serious outcome of the illness, together, represent a characteristic syndrome which to our knowledge has not been described before' (1, p. 68).

Long QT syndrome

Similar individual cases had been observed previously, and after Jervell and Lange-Nielsen's article, more began to appear. The first of these was in the following year in a case report in the *New England Journal of Medicine* (22). In 1949, Woodworth and Levine had observed a deaf-mute boy with fainting episodes and prolonged QT intervals, but their work was published subsequent to them reading Jervell and Lange-Nielsen's article. Thus, the condition was very nearly named Woodworth and Levine syndrome (23).

Patients with similar fainting episodes and prolonged QT intervals, but without deafness, were described in Italy (24) in 1963 and in Ireland in 1964 (25). This form is known as Romano-Ward syndrome. Again, history could have taken a different turn here. In July 1958, a woman in her twenties was admitted to Department 9 of Ullevål Hospital for repeated spontaneous loss of consciousness, which eventually was accompanied by seizures (26). She was examined by Fred Lange-Nielsen, a junior registrar. This was a patient with Romano-Ward syndrome. Five to six years before it was described in published research by others, the condition was documented in Ullevål's records but not fully understood. In hindsight, it is easy to think that Lange-Nielsen should have connected the dots. After all, the only thing missing compared to his and Jervell's descriptions the year before was the deafness (26). There must have been some speculation, as Lange-Nielsen wrote in the medical records that the patient's family members were examined for a suspected 'familial anomaly of an enzymatic nature'. The mother and siblings showed similar ECG changes, but these were 'less pronounced', while the findings for the father were normal. The Ullevål records do not indicate whether the patient's condition was discussed with others (26). The logical step would have been to consult Jervell,

but at the time, there was more than one floor separating Department 8 and 9, and the medical departments were saddled with psychological and territorial barriers. Unfortunately, the woman died suddenly a few months later (26).

The use of ECG equipment in hospitals started to increase in the 1940s, and quinidine, which prolongs the QT interval, was a common treatment for arrhythmias. Today, we know that the potentially fatal 'quinidine syncope' is caused by iatrogenic prolonged QT, which predisposes to heart rhythm disturbances (ventricular tachycardia). In 1963, Rolf Rokseth (1923–2009) and Ole Storstein (1909–98) at Rikshospitalet described the outcomes of 274 patients who received high-dose quinidine for the conversion of atrial fibrillation. Twelve patients experienced dramatic fainting episodes, but the authors did not mention the QT interval. This exemplifies the saying, 'You see only what you look for, you recognise only what you know' – a quote attributed to the radiologist Merrill C. Sosman (1890–1959).

The vigilance of Jervell and Lange-Nielsen led them to recognise that when several children in a sibling group were affected, there must be a hereditary link between deafness, prolonged QT intervals on the ECG and sudden death. Although they did not understand the mechanism, they knew that this was a syndrome and that the condition could be 'a possible cause of inexplicable death in children' (1, p. 68).

One reason for the major and rapid significance of the article was that it was published in the prestigious *American Heart Journal*, founded in 1925. Jervell had also previously published a case report in the journal and was appointed a member of the editorial board in 1953. Among the scholarly heavyweights from renowned institutions worldwide, it is interesting to find a name from a small town in Norway. Medical history features several pioneering descriptions from Norway that were overlooked due to limited international interest and consequently credited to others, including Spielmeyer-Vogt disease, Huntington's disease and Hallervorden-Spatz syndrome.

Significance of the discovery

Jervell and Lange-Nielsen's article is regarded as the most significant Norwegian contribution to the literature on arrhythmia (27, p. 76). The description sparked an international wave of clinical and basic research that continues to this day (28, p. 332). The 1957 article has been cited over 1400 times to date (webofscience.com, 5.8.2024). An authoritative review of Jervell and Lange-Nielsen's article from 2006 states that 'There are not many instances in medical history of a single case report so critical for the development of the subsequent knowledge on a given disease' (29).

This article was the first to document a hereditary heart rhythm disorder and confirm prolonged QT intervals as a risk factor for arrhythmias and sudden death. It opened the door to the discovery that prolonged QT intervals were a risk factor independent of the cause, and that various electrolyte imbalances

and medications could affect ion channels, leading to prolonged QT and life-threatening arrhythmias (30). It was only in the late 1980s that this was recognised as a widespread problem.

It became clear in the 1990s that there was a genetic explanation for Jervell and Lange-Nielsen syndrome. In 1991, it was discovered that the mutation was located in chromosome 11, but the gene was misidentified. The problem was finally solved in 1995 (23). In most cases, the cause is a defective potassium channel due to mutations in the KCNQ1 gene on chromosome 11 p. Otherwise, mutations in the KCNE1 gene on chromosome 21q (subtype 2) are the cause. The link between deafness and arrhythmia was finally understood. A flow of potassium ions through the cell membrane is essential for maintaining normal function in both the inner ear and the heart muscle. Mutations in the potassium channels that disrupt this ion flow result in both hearing loss and irregular heart rhythms.

A case report in 1990 provided new insights. A woman with Jervell and Lange-Nielsen syndrome gave birth to a child with Romano-Ward syndrome (31). Jervell and Lange-Nielsen syndrome follows a recessive inheritance pattern and is rare. Romano-Ward syndrome follows a dominant inheritance pattern, is much more common, milder and is not associated with deafness (2). The relationship between the syndromes was first described in 1975 (32). The same genes are affected in both syndromes, and malfunction in one type of ion channel in heart muscle cells can cause the disease. The mutations, and thus the severity of the disease, can vary between family members. It is estimated that 1 in 200 000 babies in Norway are born with Jervell and Lange-Nielsen syndrome, with four common KCNQ1 mutations present (33). In a retrospective case review, the syndrome was found to be diagnosed in 2.9 % of children in Norway with a cochlear implant (34). Deaf children being considered for cochlear implantation are therefore routinely screened with an ECG to detect the syndrome, and their heart rhythm is monitored during surgery and when the sound is activated. Today, the diagnosis is confirmed through genetic testing.

In 1957, there was no effective treatment for the syndrome, and the mortality rate was extremely high. Patients with Romano-Ward syndrome were also treated with all available antiarrhythmic drugs, but in 1975, an Italian, Peter Schwartz, demonstrated that beta-blockers were both effective and safe. Case reports and animal experiments had convinced him that the sympathetic nervous system played a key role in arrhythmogenesis. Those who experienced relapses while on beta-blockers underwent sympathetic surgery (ablation of the left stellate ganglion) (32). This remains the standard treatment, and in addition, implanted defibrillators have become a safety net following therapy failure. The mortality rate is now low.

There are long lists of medications that prolong the QT interval and that patients should avoid (crediblemeds.org), and patients now receive lifestyle advice tailored to their genotype.

Jervell, Lange-Nielsen and Kloster

Diseases and syndromes have long been given eponymous names based on who first identified them, but this tradition is contentious and is gradually being replaced by more descriptive terms that better capture the nature of the condition. Following their published work in the *American Heart Journal*, Jervell and Lange-Nielsen joined the distinguished ranks of Asbjørn Følling (1888–1973) with Følling's disease, Sigvald B. Refsum (1907–91) with Refsum's disease, and Carl Müller (1886–1983) and Francis Harbitz (1867–1950) with Müller-Harbitz disease.

It was undoubtedly Anton Jervell who was the leading expert in describing the syndrome. He was an internist with a doctorate in ECG and a special interest in cardiology (a medical specialty that was not officially established until 1965). He followed up on the syndrome with several articles. In contrast, Lange-Nielsen specialised in pulmonology and never wrote about heart disease again. For him, cardiology was a career sidetrack. In 1958, when he was faced with a similar clinical picture but without deafness, he failed to recognise it (26).

When Jervell and Lange-Nielsen wrote their article in 1956, they were 55 and 37 years old, respectively, and working as a senior consultant and a locum in Tønsberg's medical department. However, by the time the article was published the following year, both had moved on. Lange-Nielsen was undertaking a ten-month study exchange in New York, while Jervell had been appointed senior consultant and professor at Ullevål Hospital. In a way, their careers followed a similar trajectory: in addition to their time in Tønsberg, Jervell headed Department 8 at Ullevål for the final 15 years of his hospital career (8), while Lange-Nielsen worked in Department 9 for the last 25 years of his working life (16).

Johan Kloster's role in the story is intriguing (Figure 5). He had examined several members of the family in the case report and identified the prolonged QT intervals. Kloster was the same age as Jervell, and they had known each other from Ullevål in the 1930s (35). From 1940, Kloster was a senior consultant at the newly established medical department in Arendal, much like Jervell's role in Tønsberg (36). He was acknowledged at the end of the article for 'his kind assistance in placing information, electrocardiograms, etc., at our disposal', but he was not credited as a co-author and his name was not therefore associated with the syndrome. Given Lange-Nielsen's more subordinate role, the syndrome could easily have been named Jervell and Kloster syndrome instead.

Figure 5 Johan Kloster. Photo from 1960, Kjell Lund Hvoslef/Hvoslef Foto AS, AAA.PA-2425.8519, archives of Aust-Agder Museum and IKS archives, Kuben Arendal.

Jervell followed up his discovery and published five articles on the syndrome over a period of almost 30 years (1, 37-40). In 1966, he presented three new cases. Two were investigated in Trondheim and the other one at Ullevål, and the resulting article was also published in the *American Heart Journal* (37). At that point, 17 known cases had been identified worldwide. A year later, Jervell published a new case in the *Nordisk Medicin* publication (38). When he took stock in 1971, at 70 years of age, 27 cases had been reported (39). In 1985, Jervell wrote that of the eight patients he had seen himself, four had died, and among the four who had survived, two had married, one had given birth to a child with no complications and one had a driving licence (40).

In 1966, Jervell expressed a preference for calling the condition surdocardiac syndrome (surditas = deafness) (37, 41), and he used this term in his final article in 1985. Others suggested calling it cardio-auditory syndrome, but

neither term gained traction. Since the 1960s, the condition has commonly been referred to as Jervell and Lange-Nielsen syndrome.

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The article has been peer-reviewed.

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