ISSEC: An International Scientific Society of Ectopic Calcification dedicated to the advance of knowledge and awareness for ectopic calcification diseases.

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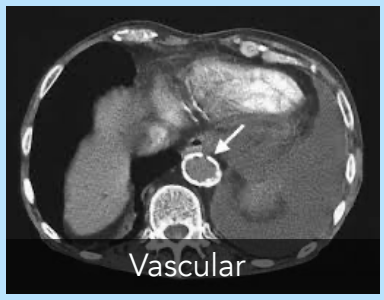
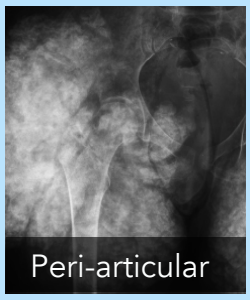
The International Society of Ectopic Calcification (ISSEC) was recently created following the successful COST Action 'EuroSoftCalcNet' that ended in 2021. Its main aim is to **facilitate contacts between all actors dealing with ectopic calcification** (EC).

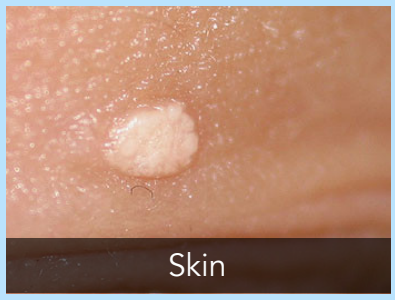
- **March 2017** — 25 countries; >200 participants
- **September 2021** — Closing Meeting (Angers) EuroSoftCalcNet
- **March 2022** — Foundation of ISSEC
- **June 2022** — **Membership applications open**

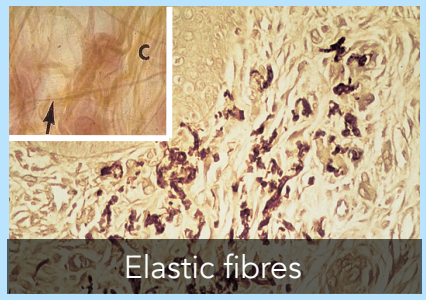
ECTOPIC CALCIFICATION

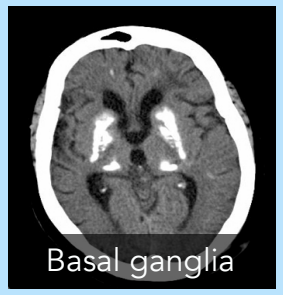
EC is defined as inappropriate biomineralization in soft tissues. Affecting a large variety of tissues such as arteries, kidneys, brain and connective tissues, EC is accelerated in **aging**, in a wide number of **hereditary diseases** as well as in **acquired diseases** such as chronic kidney disease (CKD). EC diseases often have an unpredictable evolution and are still largely incurable because of the partially uncovered mechanisms and interindividual variability. Furthermore, there is limited awareness for EC in the medical community and among the general population.





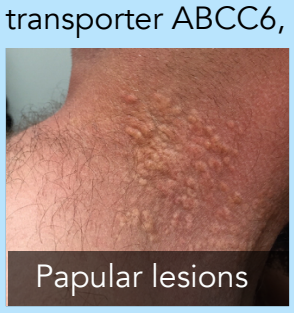


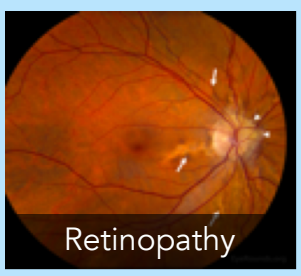


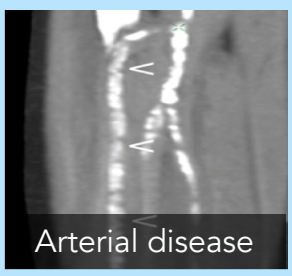


FROM RARE DISEASE TO UNDERSTANDING ECTOPIC CALCIFICATION

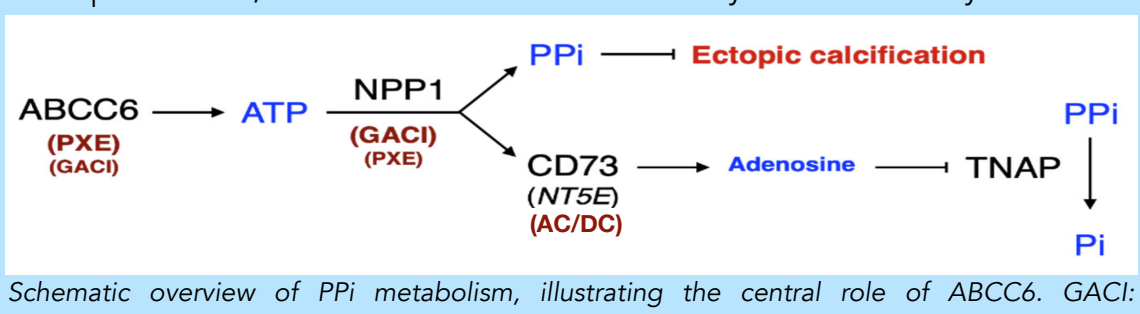
Studies of rare hereditary EC disorders are instrumental in **understanding** many aspects of **EC pathophysiology.** For example, the hallmark multisystemic EC rare disorder **Pseudoxanthoma Elasticum** (PXE), was essential for our understanding of the pivotal role of inorganic pyrophosphate (PPi) metabolism, a physiological calcification inhibitor that is significantly decreased in PXE but also in other rare disorders, e.g. GACI, and acquired diseases such as CKD. In the latter, PPi levels are inversely correlated with cardiovascular disease risk and mortality. The liver transporter ABCC6, deficient in PXE, was shown to be the main source of plasma ATP, which is then converted to PPi by the ENPP1 enzyme.





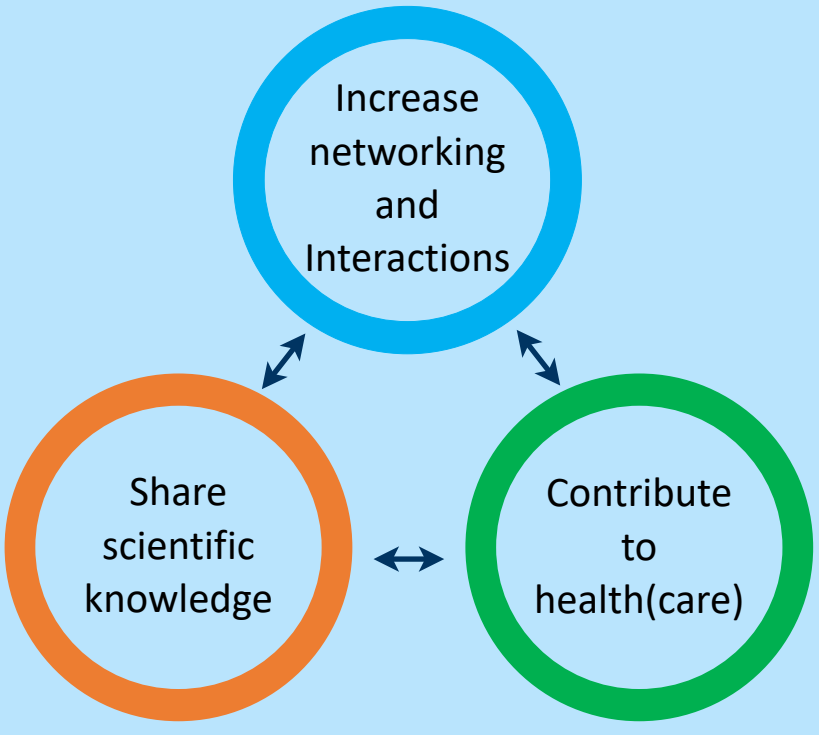


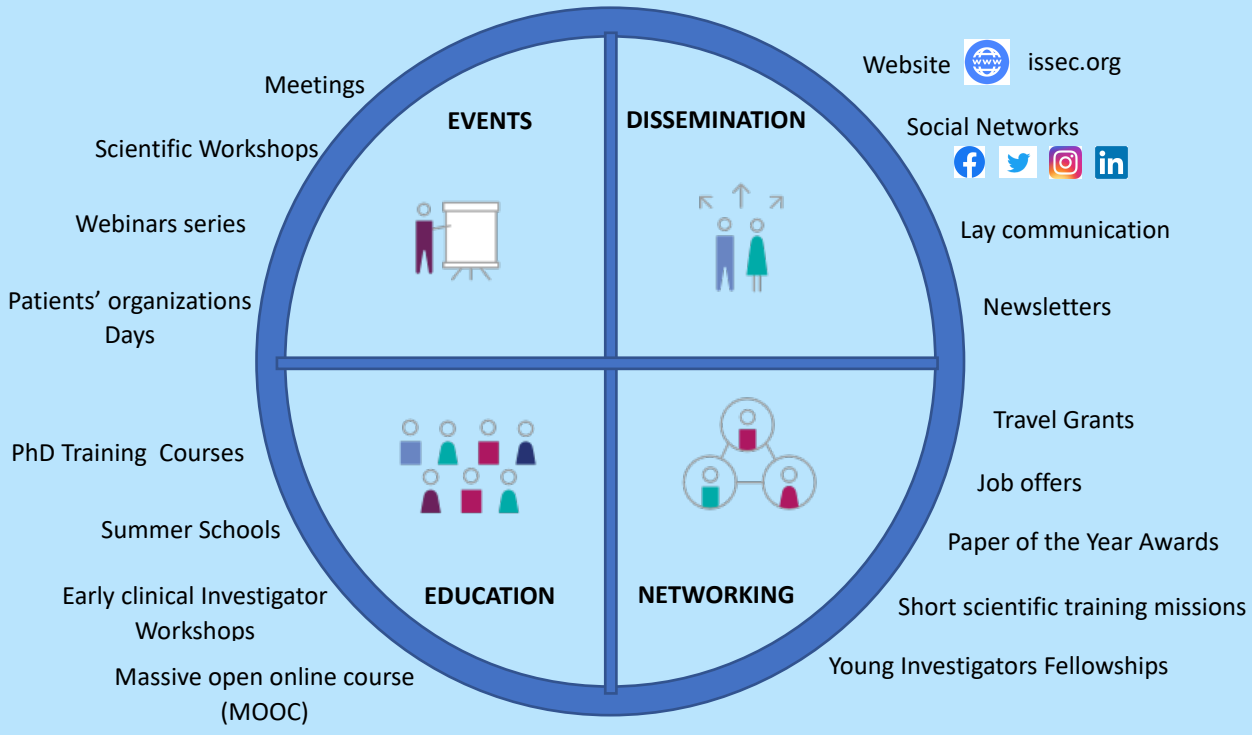
Main clinical features of PXE in the skin, eyes and the cardiovascular system



Schematic overview of PPi metabolism, illustrating the central role of ABCC6. GACI: Generalized arterial calcification of infancy; ACDC: arterial calcification due to CD73 deficiency

THE AIMS AND PLANNED ACTIVITIES OF ISSEC





By joining together all stakeholders - patients and their families, clinicians, researchers - ISSEC aims to foster **new discoveries** to **better understand**, **manage** and **treat** both rare and chronic, acquired EC disorders and to create worldwide **awareness** for the EC burden and to **improve** EC patients' position.

WANT TO KNOW MORE and/or WANT TO JOIN US?

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