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The pre-existing vulnerabilities of patients with rare disorders in Poland during a global pandemic

The global race to find a vaccine and effective treatments for COVID-19 has already shown its side effects. In the UK and Poland patients with lupus, a rare, autoimmune disease and patient organizations have raised their concerns about the availability of hydroxychloroquine and chloroquine (Boseley 2020; Godlewska 2020). In Poland, chloroquine is solely manufactured by the Adamed Group, and is marketed under the name *Arechin*. Approved as a "supplementary drug for SARS-CoV-2 infections", all of its stockpiles have been transferred to Polish hospitals (PDS 2020). As the state appropriated the available supply of *Arechin*, patients with lupus, among others, were left without lifesaving treatment. In mid-March, recognizing that the drug would most likely not "be available for individual patients in pharmacies in the near future", the Polish Dermatological Society recommended the afflicted "try to contact a specialist or POZ [Primary Care] in order to establish [a course of] alternative treatment" (PDS 2020). Like their counterparts in the UK, Polish patients were deprived of needed medication. In the words of a long-term user of *Arechin*: "We won't be killed by COVID-19, but by lupus!" (Godlewska 2020; see also Boseley 2020). After I finished writing the first version of this text, however, the Polish Dermatological Society announced on its website in early April that *Arechin* and *Plaquenil* (hydroxychloroquine) will eventually be available in restricted amounts (60 pills per patient, per month) for patients with lupus and other autoimmune diseases in the upcoming days. No further announcements about the real availability of these drugs followed.

Arechin users belong to the minority of people with rare disorders for whom any drug is actually available. For the over 6000 rare disorders, a mere 160 orphan medicines are authorized by the European Commission (EMA 2020). Still, as I show

below, the current pandemic affects individuals with rare diseases and their families in a number of ways. Whereas the WHO reassures us that in most cases the virus will only cause "mild to moderate respiratory illness," it nonetheless also warns that along with older people, "those with underlying medical problems (...) are more likely to develop serious illness" (WHO 2020). In this view, the elderly and those with chronic and rare diseases are both more susceptible to the virus and in danger of experiencing it more severely. A sole focus on COVID-19-related vulnerability obscures the issue that it is precisely those considered "most vulnerable" (Turner 2019) who are affected by the current pandemic, even if they do not contract the coronavirus itself. As Vincanne Adams (2020) aptly puts it, "the turn to pandemics displaces other forms of healthcare for ongoing morbidities (...). [L]ike other disasters, the actual arrival of COVID-19 magnifies pre-existing vulnerability." In this contribution I show that privileging COVID-19-related vulnerability disrupts the delivery of care for people with rare metabolic disorders, who were already structurally vulnerable (Quesada et al. 2011).

A multivalent concept, vulnerability in anthropology, as Marja-Liisa Honkasalo (2018: 8) reminds us, "has mainly been 'good and dark', a descriptive moral term for describing the quality of life and the condition of the poor and excluded." In philosophy, vulnerability has increasingly been used to denote the universal human condition, "our shared vulnerability" (Cole 2016: 274; see also Honkasalo 2018; Mackenzie 2013). Critical of this approach for its inability to address systemic inequalities, Alyson Cole among others emphasized that "this is a structural matter as well as an experiential one. People are subjected to or immunized from vulnerability in radically distinct, different and unequal ways" (2016: 265).

Positioning lies at the core of the concept of structural vulnerabilities that has become an important conceptual tool in medical anthropology (Quesada et al. 2011; Castañeda 2013). According to James Quesada and his colleagues, "the vulnerability of an individual is produced by his or her location in a hierarchical social order and

its diverse networks of power relationships and effects" (2011: 341). They further highlight that structural vulnerability "focuses on how a host of mutually reinforcing insults (ranging from the economic and political to the cultural and psychodynamic) that dispose individuals and communities toward ill health are embodied" (Quesada et al. 2011: 344).

Areas of structural vulnerability

My research team[1] is conducting comparative ethnographic research with people suffering from rare metabolic disorders, such as fatty acid oxidation disorders and organic acidurias as well as their families and professionals in Poland, Finland, and Sweden. A special and restricted diet is the only available "medication" for such conditions. The treatment itself is clinically challenging and it subjects patients and their family members to a dietary regimen for the rest of their lives. This regime is based on so called "foods for special medical purposes" and requires frequent check-ups with a variety of specialists; during the current pandemic, adhering to both has become even more challenging than usual.

A display stand of Nutricia, producer of a number of special foods for people with rare metabolic disorders among others. The slogan in the background reads in Polish: "We help. We inspire. We are." The picture was taken at a rare disease conference in Sekocin near Warsaw in November 2016. Copyright: M. Rajtar

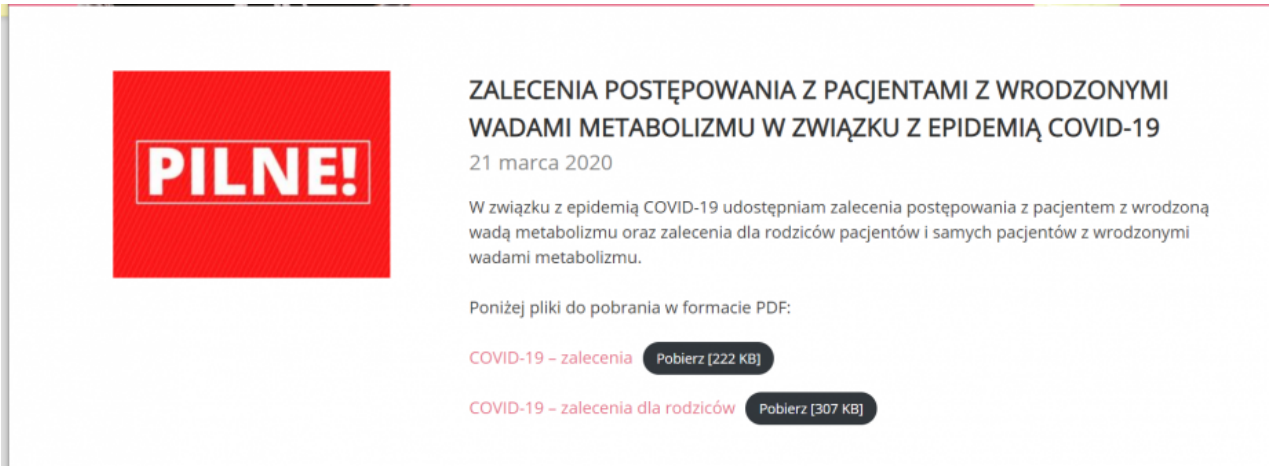
First, in some fatty acid oxidation disorders, two powdered formulas, i.e. Lipistart® (Vitaflo/Nestlé) and/or Monogen® (Nutricia/Danone), are considered essential. Along with another important "food product"– MCT (medium-chain triglyceride) oil – these formulas are available on prescription, imported via pharmacies and reimbursed by the Ministry of Health. However, as access to these products is restricted to a six-month supply/patient in Poland, patients, their parents, and doctors are required to regularly apply in writing to the Ministry of Health for

reimbursement. Overwhelmed by bureaucracy and delays on a regular basis, access to essential treatment may be hindered by the current COVID-19 pandemic even further. Such concerns are raised by some parents who are active on Facebook (FB) support groups. In Poland such groups are comprised of 80 to 800 members, mostly parents of children with rare metabolic diseases and patients. For instance, one mother wrote "What about our kids? What about their supplies?" (notes by Filip Rogalski, researcher in the project, 1/04/2020). Her emotional appeal was answered by others who claimed that their requests for imports and reimbursement were quickly processed or actually confirmed that the bureaucracy took longer than usual.

Second, as a number of parents have shared on the same support group, check-ups have lost their regular rhythm; they have either been cancelled or they may be cancelled at any time, thus parents fear for the wellbeing of their children. This was confirmed by a dietitian who works at a leading specialized medical center in Warsaw (personal communication, 2/04/2020). This fear is compounded with that of the possibility of even contracting the coronavirus when going to the hospital. Admitting that for Rare Inherited Metabolic Diseases (IMDs) patients and their caregivers the "fear of being infected with the virus" is "normal", the MetabERN (European Reference Network for Hereditary Metabolic Disorders) recommends "NOT to interrupt or postponed [sic] your therapy unless it has been directly recommended by your metabolic expert or become inaccessible/unavailable due to the current strain put on health services by the COVID-19 pandemic" (MetabERN 2020; capitalized in original; original spelling). In other words, in times of pandemic, the agency of structurally vulnerable patients and their caregivers is more limited than ever (Quesada et al. 2011: 342).

Furthermore, many patients who are afflicted by rare metabolic disorders are prone to acute metabolic decompensation that may be triggered by infections, physical activity, fasting, and stress among others. In order to prevent neurologic injury and/or death, prompt recognition of symptoms is crucial along with referral to a

specialized center, often located at a university hospital. Recognizing the danger posed by COVID-19 infection to such patients and their "high risk status," the National Consultant in Pediatric Metabolism in Poland issued recommendations that emphasized the "unconditional isolation" of both patients and their families as the best preventive method (Sykut-Cegielska 2020). These recommendations have been forwarded by a large Polish patient organization for people with PKU (phenylketonuria) and other IMDs as well as by a few support groups on FB. Some parents had already received these recommendations from their kid's doctors; others, however, were surprised to hear about their child's "high risk status" and some were simply grateful for the advice. In her recommendations, the National Consultant urges "vigilance"; she further warns that "if a child requires hospitalization, s/he may be placed in a different [medical] center than usual, i.e. at a hospital for infectious diseases" (Sykut-Cegielska 2020). This may sound prudent; however, it is imaginable that hospital staff, already overwhelmed and unfamiliar with rare conditions, would focus on treating the virus rather than acute metabolic decompensation. Parents express such concerns on FB support groups. They also complain that the information about whether parents can be present at their child's bedside or not is contradictory. In the words of one mother "*it's known that children afflicted by additional conditions (in particular rare [conditions]) require special care. I'm quite worried whether an infectious disease ward has additional knowledge about a given rare disease. Hence, it's important for a parent to be present [there]*" (notes by Katarzyna E. Król, researcher in the project, 2/04/2020)



An edited screenshot of the recommendations issued by the National Consultant in Pediatric Metabolism in Poland. The icon in red reads "Urgent!"; the following text reads: "Recommendations on Dealing with Patients with Inborn Errors of Metabolism in Regard of COVID-19 Epidemic." Source: http://pediatriametaboliczna.pl/category/aktualnosci/ Last access: 04/05/2020.

Finally, a politically and epidemiologically driven focus on the COVID-19 testing may adversely impact conducting other tests, such as follow-up tests for a positive newborn screen. This seems to be the case for at least one parent who complained on one online support group that due to the pandemic, the results of genetic tests that were conducted to confirm a condition in their newborn have been significantly delayed, thus intensifying the ambiguity of a "patient-in-waiting" position of the newborn (Timmermans & Buchbinder 2010).

A playground in Warsaw closed due to the pandemic. Copyright: M. Rajtar

Foregrounding universal vulnerability

In research ethics as well as social and public health policies, the concept of vulnerability is most often used to label specific individuals or groups who are at increased risk of or susceptibility to harm (e.g. Boldt 2019; Rogers 2013; see Turner 2019 for humanitarianism). In this view as well as in popular discourse, vulnerability is understood negatively as dependence, victimhood, and helplessness, and thus calls for extra support and/or specific intervention. This labelling conception of vulnerability is criticized for being potentially patronizing and stigmatizing. Like the universalizing approach to vulnerability espoused by a number of philosophers,

labeling individuals and groups as “vulnerable” does not acknowledge the complexity of human vulnerability; rather, vulnerability is context and case related (Rogers 2013: 70).

The current pandemic reveals our inherent vulnerability to the novel coronavirus. Although the WHO and health experts are concerned about the vulnerability of some groups, such as the elderly and people with chronic conditions, practical measures to address the virus outbreak employed by some governments seem to focus on universal vulnerability rather than that of a particular group. Appropriation of *Arechin* stockpiles for the general population in Poland neglects those for whom the drug constitutes lifesaving treatment; restricting hospital access to exclusively COVID-19 victims further disrupts the delivery of care for already structurally vulnerable patients with rare disorders; thus, even at the level of universal vulnerability, we are not equal.

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Footnotes

[1] The research team includes sociocultural anthropologists (Katarzyna E. Król, Filip Rogalski, PhD, and myself), and a doctoral student in cultural studies (Jan Frydrych). We also collaborate with a dietitian (Ewa Ehmke vel Emczyńska-Seliga). I would like to thank everybody for their contribution to the project.