SWISS SOCIETY OF NEONATOLOGY

When the Millingville Bank doesn’t crash

June 2010

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This girl had been born at term after an uneventful pregnancy more than 10 years ago in Macedonia. At the age of 7 weeks, the infant was brought to our institution because of cyanosis and failure to thrive. Physical examination revealed a respiratory rate of 60 breaths per minute and a transcutaneous oxygen saturation of 50%. The heart rate was 140 beats per minute, the heart sounds were displaced to the right and there was a 3/6 systolic murmur. There was polycythemia with a hemoglobin concentration of 185 g/l.

Echocardiography revealed complex congenital heart disease consisting of dextrocardia, a functionally univentricular heart with a hypoplastic left ventricle, a large ventricular septal defect, D-transposition of the great vessels and atresia of the pulmonary artery (Fig. 1). Due to severely hypoplastic pulmonary vessels the construction of an aortopulmonary anastomosis was judged to be impossible. At that time, the following statement was documented in her medical record: "…. The parents were informed that their child suffered from an inoperable cardiac defect and that she will die. The time of survival is estimated to be several weeks, not exceeding 1 to 2 months based on the severe and progressive cyanosis." But things turned out differently.

At the age of 7 months, a Blalock-Taussing-Shunt was established. Surgical revision was necessary one month later due to persisting hydrothorax. At the age of 11 months, pulmonary tuberculosis was diagnosed and treated (Fig. 2). At the age of 2 years, a bidirectional cavopulmonary anastomosis was performed. At the age of 4 years, she was able to walk freely but had severe neurodevelopmental delay. At that time, a Fontan procedure was discussed but subsequently rejected due to the hypoplasia of the pulmonary arteries. Instead, stents were implanted into both pulmonary arteries.

Over the past 5 years, she was hospitalized several times due to both viral and bacterial pulmonary infections. At the time of this writing she is 10 years old. The last echocardiography showed stable but reduced biventricular contractility and significant tricuspid regurgitation. Her transcutaneous oxygen saturation is around 65%. The most recent CXR is shown in Fig. 3.

Fig. 1

Babygram at the age of 7 weeks.

Fig. 2

CXR at the age of 11 months with pulmonary tuberculosis involving the right upper lobe.

Fig. 3

Most current CXR (ap and lateral views) of the patient at the age of 10 years.

DISCUSSION

Prophecy stems from the Greek word propheteia, which literally means „to speak forth“ (pro, „forth“; phemi „to speak“). The part phemi - to speak - is neutral and does not in itself imply accuracy. In contrast, the word prognosis means „to know forth“ (pro, “forth”; gignoskein “to know”). The part gignoskein - to know - implies that a prognosis is more reliable than a prophecy.

The knowing-part of medical prognostication is mainly based on population studies of at risk individuals. Predictive models accounting for multiple risk factors exist for several diseases. They form the basis for a more precise prognosis for a specific patient. For example, according to the NICHD database, the outcome of an infant born at the limit of viability is more accurately predicted by a model taking into account gestational age, birth weight, sex, prenatal steroids and singleton versus multiple birth than by gestational age alone (1).

Although the NICHD algorithm improves outcome estimation, D'Angio and Mercurio have warned of the illusion of accuracy. They explain in their article that even this model is not only inaccurate but also imprecise (2): Several input variables may be inaccurate. Obstetrical estimates of fetal weight and gestational age are often difficult to pinpoint within more than one increment of the model (100 grams and 1 week, respectively). But some variables are also imprecise. For instance, the outcome early in a week of gestation is different from the outcome late in the same week. The model is not designed to provide predictions using such a detailed scale.

In practice, even the best statistical model is only of limited value in establishing a medical prognosis. A 50% mortality rate, for example, may exist for a group of patients, but for an individual patient, the mortality is either 0% or 100%. This phenomenon is called **„hiatus theoreticus“** and is one reason for prognostic inaccuracy (3).

Another concern regarding outcome data is the risk of the so-called **self-fulfilling** prophecy. The sociologist Robert Merton introduced this concept in the 1950s. The concept of the self-fulfilling prophecy stems from the Thomas theorem (formulated by W. I. Thomas (1863–1947) in the year 1928) which states that „If men define situations as real, they are real in their consequences.“ A self-fulfilling prophecy is a prediction that directly or indirectly causes itself to become true, by the very terms of the prophecy itself, due to positive feedback between belief and behavior. The paradigmatic example is that of the Last National Bank, also known by its owner's name, the Millingville Bank: Groundless rumors of an imminent bank collapse lead to a large number of people withdrawing their savings. This in turn lead to the actual collapse of the bank.

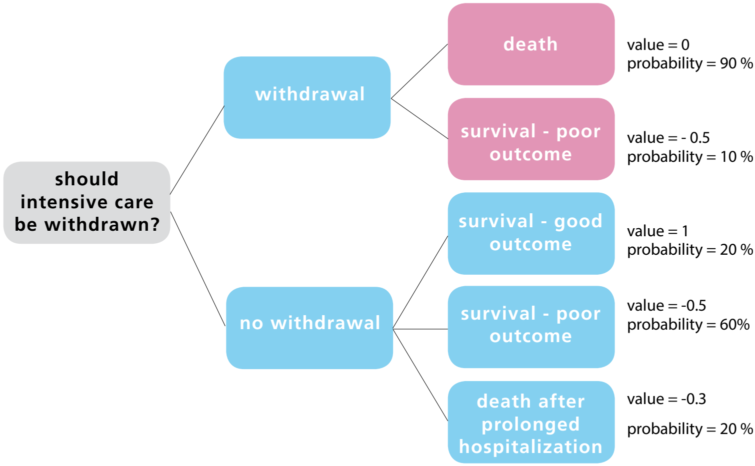
Where predictions affect outcome, it can be extremely difficult to ascertain the mortality rate for patients if all treatment were provided (4). Mercurio, for example, argues that the self-fulfilling prophecy may contribute to mortality rates of extremely premature infants. He points out that a center that has never had a survivor below a certain gestational age may deem it reasonable to refuse resuscitation below that age, as survival appears extremely unlikely or impossible. Over time, they will continue to have 0% survival rate and the perceived justification for refusal will persist (5).

In the past, prior to the advent of effective therapies, prognostication was the major task of the medical profession: the doctors were paid for their opinions. Several studies have addressed the ability of physicians to predict the future. Their results were uniformly sobering. In one study, physicians were no better than chance at identifying patients who would survive in-hospital cardiopulmonary resuscitation (6). At the delivery of extremely premature infants some neonatologists rely on “how the baby looks” when deciding whether or not to initiate resuscitation. It has been shown that observers’ ability to predict survival was poor (AUC 0.61, 95% CI: 0.55-0.67 at 5 minutes of life). Interestingly, the level of experience did not affect the observers’ accuracy of predicting survival (7). Christakis et al. have found that prognostic accuracy decreased in terminally ill patients as the duration of the doctor-patient relationship increased (8). Overall, the conclusion is clear: doctors are bad prophets.

Even the most refined predictive models and the most experienced physicians are not able to prognosticate with a 100% accuracy. Prognosis refers to the future and always contains an element of uncertainty. This has great implications for medical treatment limitations as prognostication is the fundamental basis for the determination of futility.

One approach to decision-making under uncertainty is to use the **utility** theory of Von Neumann-Morganstern (9). It has its origin in economics and deals with the analysis of choices within risky projects but can also be adapted for decision-making in medicine. The probabilities of different outcomes are combined with the values attached to those different outcomes to determine which course of action will lead to the greatest expected utility. By convention in medical decision-making, life in full health is assigned a value of one, and death a value of zero. Outcomes that are judged to be worse than death are given negative values. However, there are different philosophical opinions about whether life with severe impairment is worse than death. An example of using decision theory is shown in Fig. 4.

Decision tree: **Should intensive care be continued or withdrawn in a certain patient?**



Equation: **Expected Utility (A) = Probability (x)*Value (x) + Probability (y)*Value (y)**
Expected Utility (withdrawal of intensive care) = 90 %*(0) + 10 %*(-0.5) = - 0.05
Expected Utility (no withdrawal of intensive care) = 20 %*(1) + 60 %*(-0.5) + 20%*(-0.3) = -0.14

In this example withdrawal of intensive care has the greater expected utility and is the preferred action.

Fig. 4

Example using the utility theory by Neumann-Morgan-stern.

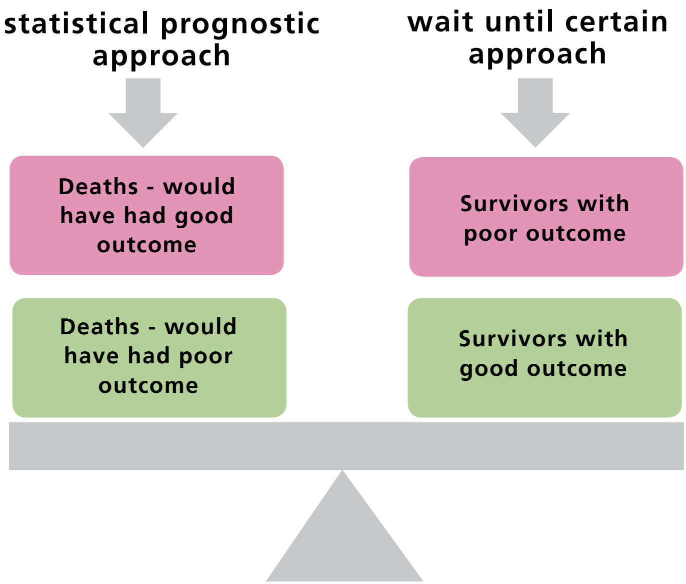


Fig. 5

The “wait until certain” strategy increases the number of survivors with bad outcomes. The “statistical prognostic” approach increases deaths that would have had a good outcome.

Applying the utility theory to clinical situations seems to be quite difficult. In her article “Treating Baby Doe”, Rhoden describes different practical approaches when medical prognosis is uncertain (10). At one extreme, there is the **„wait until certainty“** strategy consisting of continuing aggressive treatment until death or irreversible coma has become certain. It minimizes the risk of not treating an infant who would have been treated had doctors known the (positive) outcome; but it also maximizes the possibility of saving an infant who would not have been treated had doctors known the (negative) outcome. This principle ignores statistical probabilities. At the opposite side of the spectrum is the **“statistical prognostic”** approach: it withholds treatment from infants for whom the statistical prognosis is considered to be grim. This approach minimizes the number of infants who survive with profound handicaps but accepts to sacrifice some infants with a potentially good outcome (Fig. 5). This approach, of course, is only as good as the statistical data upon which it relies and may lead to self-fulfilling prophecies. The strategy in between is called “individualized prognostic” approach: Treatment is started and frequently reevaluated. It takes individual factors and the caregivers’ opinion into account and acknowledges that there is always some degree of prognostic inaccuracy.

The case described above is an example of an unexpected survival. The prognosis was felt to be grim and incompatible with life. But the child survived the neonatal period, was cured form pulmonary tuberculosis and is know 10 years old. Prognostication is an important part of the medical art but, as this example illustrates, some prognoses resemble prophecies - and prophecies may not come true.

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