

Frequency Of The Congenital Defect-Anomalies Of The Extremities In The Federal Republic Of Germany*

By PROF. DR. OSCAR HEPP

*Director, the Orthopedic University Clinic and Polyclinic,
(Hueffer Foundation), Muenster, Federal Republic of Germany*

A survey of the recent increase in frequency of phocomelia based on the number of cases observed at the University Clinic and Polyclinic, Muenster, up to the end of 1961, is contained in this report. Advice to be given to the parents of afflicted children is outlined; and steps necessary for the prosthetic care of these children in order to prepare them, as far as possible, for school and a profession are also presented.

In the last two years the number of children born with severe congenital deformities of the extremities has risen by leaps and bounds in the Federal Republic of Germany. Already, the damage done is so catastrophic as to require a service of extraordinary magnitude. While the full extent of the damage cannot be estimated as yet, the known number of afflicted children is already so large that one can predict which measures will be necessary and in what manner the many parents concerned should be advised.

Based on the number of children examined at the Orthopedic University Clinic and Polyclinic up to the end of 1961, this study provides the first survey as to the kind and frequency of the present extremity deformities which, in combination with other anomalies, presents a situation far exceeding other previous experience. Types of congenital extremity deformities once rarely seen are now very prevalent and constitute a much higher proportion of the total population.

The opinion has been frequently expressed that the anomalies were caused during the first weeks of pregnancy by one or more drugs or other substances which were put on the market in the Federal Republic in recent years and used extensively. This view is probably correct, but has not yet been proven in detail. It is not appropriate for me to question the causes in this study, since many others are now working feverishly to determine the possible causes.

However, it appears certain that our initial presumption that the anomalies were caused by radioactive fallout from atomic bomb tests was incorrect. No similar accumulation of severe extremity deformities have been observed in the fallout regions of the rest of the world. Thus we are apparently confronted with some unknown factor(s) X, Y or Z as toxic agent(s).

Table One Discussion

Table 1 indicates the extent of the damage among the respective patients in our clinics. The number of children with extremity anomalies is shown in squares per case according to year of birth. Anomalies of the extremities

* Reprinted by permission of the author and publisher from *Inter-Clinic Information Bulletin*, July-August, 1962.

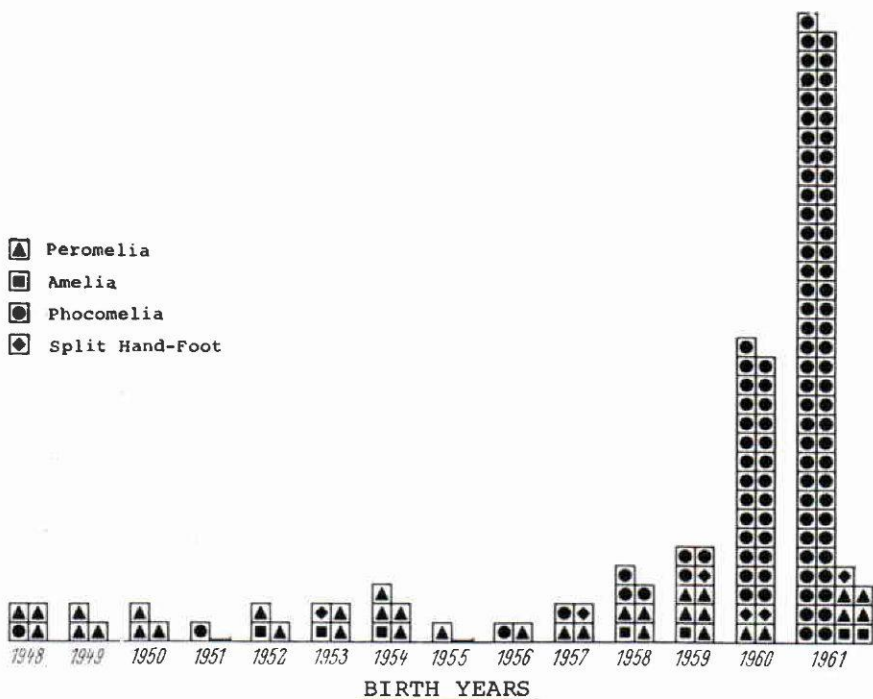


TABLE ONE 148 DEFECT-ANOMALIES OF THE EXTREMITIES

have been registered, but other orthopedic conditions such as clubfoot, dysplasia of the hip joints, arthrogryposis multiplex, vertebra deformity, etc., are not shown. A triangle represents a peromelia: a defect of the extremity which resembles an amputation, even though bud-like, rudimentary parts of the peripheral limb segment sometimes exist in great variety. A square represents amelia; complete absence of an extremity at shoulder or pelvic girdle. A circle represents phocomelia: seal-hands or stump-limbs of various lengths with predominant defects of the medial limb part and of the thumb side. A rhombus represents peripheral hypoplasia of the type involving split hands and split feet.

Only seven cases of these types of extremity deformities were seen in our clinic during the individual birth years of 1948-58. Of these, the quasi-amputations, amelia and peromelia were predominant. Since 1960, however, a large number of phocomelia extremity stumps, which had appeared only ten times between 1948-1959, with six in 1958-1959 alone, suddenly appeared. Among our 148 cases of extremity deformities, there were 27 cases of phocomelia in 1960 and 65 in 1961.

The avalanche of phocomelias is thus recognizable at a glance. The climax of 1961 may not have been reached at this time, since many of the babies may not yet have been presented for clinical consultation. The increase between 1960 and 1961 is therefore probably considerably higher. What the birth year of 1962 will show cannot be anticipated. Even if a drug is found to be the cause and eliminated from the market, figures similar to 1961 are expected.

The unique rise of phocomelia indicates that something absolutely new has occurred. One cannot assume that perhaps the research and development

efforts of our clinic in the field of fabricating prostheses was responsible for the increased admittance of children with extremity deficiencies since other clinics have shown similar increases.

One hundred and twenty-five of our cases are in the Nordrhein-Westfalen region. Ninety similar cases who were seen by Professor Imhaeuser in the Department of Orthopedics of the City Hospital in Dortmund, and numerous children who were presented in Wuppertal, Dusseldorf, Koeln and Bonn, should also be added to the total, and new patients are being presented daily in increasing numbers as a result of instructions given to the State Health Departments in the reporting of such cases.

Consideration of the files of Dr. Pfeifer of the University Children's Clinic in Muenster and of Dr. Herbig, State Physician for Disabled from Westfalen-Lippe, plus the records of our own clinic, indicate there are more than 200 children with defect anomalies of the extremities at the present time. A total of 1,000 cases in the North German region may be a conservative estimate.

Table Two Discussion

Table 1, as stated earlier, indicates the frequency of the cases and their distribution between four kinds of deformities, with a rising increase in phocomelia cases since 1960. Table 2 represents the distribution of extremity involvements in the 148 cases. The black corner of the case-square represents a malformation, and according to the direction, shows left or right, upper or lower extremity. In this Table, the severity of the malformations in each of the four groups is subdivided in order to demonstrate the magnitude of the rehabilitation task in general and for the affected individuals.

As noted in Table 1, cases of amelia, peromelia, and split hands and feet have been observed in a fairly even proportion over the years. Thus in Table 2, our attention is focussed on the forms of phocomelia which have recently appeared in such large numbers.

The most frequent cases are those with seal hands in which a more or less complete elbow joint is inserted in the intercalary piece. Occurring

Peromelia			Amelia	Phocomelia				Peripheral Hypoplasia
<div><div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</div><div>8</</div></div></div>								

Explanation of Symbols: Defect of Extremities

◇ One Person ◇ Upper ◇ Double ◇ Lower ◇ Double ◇ Minor Degree

TABLE TWO 148 DEFECT-ANOMALIES OF THE EXTREMITIES

almost as frequently are those cases with seal hands and short intercalary pieces, followed by the seal hand and long intercalary piece group, both of these being without an elbow joint. All three of these predominant subdivisions are distinguished by the absence of the thumb and in longer extremities by more or less large radius defect. They consistently lead to some cases of club hands without thumb with or without partial radius defect, and club feet with fibula defect, partly also with tibia defect.

Therefore, we have intentionally included as a group of the phocomelia deformities those cases with loss of thumb and a radius or fibula defect, which is usually considered a special group of anomalies. The symmetric bilateral occurrence of these conditions in the upper extremities of the recent birth groups indicates clearly that this deformity pattern should also be evaluated as a reduced form of phocomelia.

Cases of seal hands with complete absence of an intercalary piece occurred less frequently than those involving an intercalary segment. However, this group of twelve patients included four with quadrilateral defects.

The number of cases characterized by a bud extremity, or individual finger buds at the shoulder girdle, were least frequent of all and verge on the amelia classification.

There is no clear distinction between the peromelia and the phocomelia groups. In our classifications we had to determine whether a limb defect more greatly affected the middle part of the limb or whether the distal part of the limb was so severely underdeveloped that an amputation-like condition obtained. In our classification, therefore, we knowingly placed the amelia between the peromelia and the phocomelia because there are consistent transitions to either side.

It would be difficult to specify the multitude of variations, especially since the transitory forms are less common than the typical phocomelia cases. Furthermore, a study of the data reveals that the upper extremities are afflicted more frequently than the lower, and that only two cases of phocomelia of both legs alone (club feet with short intercalary pieces), were found in our series. Among the total number of 102 cases of phocomelia, there were just seven cases in which one or both legs alone were involved. The arms were deformed in 95 cases—only three times unilaterally.

Among the 92 arm phocomelia cases, the left side was shorter and afflicted more severely than the right side on six occasions. The right side was more deformed than the left in only two cases. It has been known for a long time that there is a preponderance of left side deformities among the amputation-like below-elbow peromelia cases. Our data reveal thirteen left side peromelia cases, compared with nine right side cases.

A pathetic sight is presented by nine children with phocomelia of all four extremities, five with short deformities, four with longer ones; and five others with triple deformities (only one leg not afflicted).

Comparison With U. S. Cases

The impact of the current crisis in West Germany is most strikingly illustrated by a comparison of the 148 classified cases of defective extremities at our clinic in Muenster with the distribution of extremity in 273 children compiled at the Michigan Crippled Children Commission's Clinic.

The fact that the types of deformities, despite a difference in classifications, were divided and sub-divided into the same groups according to photographs, as our own cases, greatly aided the comparison. Although the American series was only twice as large as the Muenster sample, their data shows approximately five times as many peromelia and amelia cases

as ours; on the other hand, the ratio of phocomelia cases in the two samples is approximately 1:1 despite the difference in total numbers, with the Muenster upper extremity sub-group outnumbering their American counterparts by approximately 3:1.

The differences in the two samples are even more clearly illustrated by a comparison of the percentage of total cases falling in the major categories of defects. The pertinent figures are:

	Muenster	USA
Peromelia-Amelia	25%	70%
Phocomelia		
Upper Extremity	62%	11%
Lower Extremity	13%	19%

(Frantz, Charles H., M.D., and O'Rahilly, Ronan, M.D., "Congenital Skeletal Limb Deficiencies", *The Journal of Bone and Joint Surgery*, 43-A, 8, December 1961.)

The increased frequency of upper extremity symmetrical (bilateral) phocomelia cases is perhaps best indicated by the following: the relation of the bilateral to the unilateral cases in our phocomelia group is in the proportion of approximately 5:1, compared to the United States ratio of 1:3.

Besides the extremity deformities, the present syndrome frequently shows haemangioma, especially on the forehead and upper lip, saddle-nose, deformities of the external, perhaps also of the inner ear, stomach—intestinal atresia and heart deformities. In the event that the latter two are severe, the children die early or cannot live at all. Only an accurate study of the still-born and of abortions would reveal the total embryopathy caused by one or more toxic factors.

Other deformities of the skin, the ear and the internal organs have not yet been surveyed. However, a careful pediatric and otological examination is indicated even if the superficial symptoms do not imply circulatory and nutritional disorders.

In the last two birth years, hydrocephalus was noted once in a child with extremity deformities. A marked mental defect was also noted once during this period. However, most children have alert, pretty and kind expressions. Ear deformities and haemangioma are, of course, disfiguring, but intelligence does not appear to be affected. A three-year-old girl with a four-fold deformity (bilateral upper extremity amelia, phocomelia with a long intercalary piece at the right leg and peripheral phocomelia of the left leg) is advanced for her age in intelligence and development and is the special favorite of the ward.

Inquiries to colleagues in the countries bordering Germany—e.g., Denmark, England, Holland, Belgium, France, Switzerland, Australia and in the Soviet occupied areas of Germany, revealed that no special increase in frequency of phocomelia cases has been observed. Several cases in the vicinity of Basel and in East Berlin, however, indicate a spreading of the toxic agent across the border. Very recently we learned of an accumulation of embryopathic cases in England and Australia, from areas into which one of the drugs in question has been exported in sizable quantities until recently.

Immediate Action Required

This catastrophe in the Federal Republic requires that immediate steps be taken to limit the damage or to eliminate it. The population, the physicians, and the authorities should act immediately, without red tape.

1. Husbands and wives, in the event that a pregnancy is possible or probable, should avoid all drugs and possibly toxic new preparations which

have been on the market for only the last three years. Preferably, they should use old, proven drugs, detergents, etc., until the toxic agent has been correctly identified.

2. Physicians should prescribe preparations placed on the market within the last three years for only as long a period as is necessary and with utmost reluctance until factor X, Y or Z has been established as the certain cause of the anomaly-rate. Information about possible damage before and during early pregnancy is urgently required for patients in the child-bearing age. Even if the probable cause of the catastrophe should be limited eventually to one toxic factor, the public should be made aware of the danger of the thoughtless misuse of drugs.

3. Pregnancy tests with hormone tablets should be halted until the harmlessness of the tests alone or in combination with other medication has been proved. The AZR at present yields sufficient and harmless possibilities for proof of pregnancy.

4. All known possible causes of damage before and during pregnancy should be avoided, such as X-ray examinations, treatment with radio-active preparations, or with the proto-plasma poisons from the group of the zytostatica. If X-ray or radio-active treatments are essential for a woman's health, she should avoid pregnancy for an adequate period of time. Hormone treatments and agents promoting menses in treatment of an undefined amenorrhea should involve only drugs which have proven to be safe for many years.

5. Physicians and authorities should cooperate closely in order to discover the causal factors as rapidly as possible. Laboratories of the chemical industry and universities are already cooperating vigorously on this problem.

Physicians and authorities should report all cases of abortions and stillborn babies, as well as normal births with corresponding anomalies. It is my opinion that when a critical situation exists, such as the present one, there is an obligation to report without consideration of the fundamental law, since common welfare requires it for protection of future generations.

We are confronted with a new and terrible situation in which physicians and the chemical industry are concerned about their liability. This concern is unjustified until a substance in continuous use is proven harmful or if it is found that a substance has been marketed without thorough prior research regarding toxicity. Neither condition has been demonstrated to date.

The five suggestions listed above are designed to limit the damage, but emergency measures are also necessary to ease the distress of patients and parents. The number of children with severe extremity deformities, especially of the arms, is so great that insufficient institutions are available to train the children to attain their full possibilities as valuable members of the community.

Children up to the age of six and seven years, with normal intelligence and character, should be prepared for general or special schools. The development of their mental abilities is of great importance, since they will be unable to perform many manual activities during their life spans. Many of them will only become independent of care at great cost.

Parental Consultation

The immediate steps for relieving the psychic and physical distress of the parents and children concerned are as follows:

6. The parents should be reasonably and carefully informed about the future situation of their child. They should be advised of the proper role

they must play in the care of the child. The first thoughtless, ill-advised statement to parents or nursing personnel in the presence of the parents sometimes decides whether the child receives the essential "nest-warmth" of an understanding home, or whether he will become an outcast left to the care of the community.

The parental consultation should include the following:

a. The parents are not to be blamed; nor should they experience guilt feelings; or feel that they have been subjected to a "Godly punishment for sins committed." The poisoning of the pregnancy was an outside event unforeseeable up to now.

b. As far as we know at this time, these children are amiable individuals unimpaired from the standpoints of character and mental ability. Character and spirit will determine their fate much more than the defect of their extremities and internal organs (insofar as the defect will not shorten their life expectancy).

c. Character, spirit and personality have enabled people without arms to become painters of artistic accomplishment and were vital factors in helping handicapped people in other professions to lead full, useful lives.

d. In the age of automatization technical aids of all kinds are now available or can be manufactured in order to secure a full life for these children, according to their abilities.

e. If the phocomelous arms in bilateral cases have the length to touch each other at the ends, grasping functions will probably exist to make eating, writing and a large part of body care possible without technical help.

f. In extremely short bilateral arm stumps the feet will be able to take over the function of the hands in many cases, even if they are malformed but have sufficient leg length.

g. In the initial stages the children should be cared for, reared and handled in the same manner as all other babies and small children. Their extremity stumps should be activated in order to attain efficient development for future functions. Their shape is not of consequence.

h. Splints and bandages have no positive influence on growth but rather hinder activity and functional development.

i. These children grow up according to their mental ability in the same rhythm as normal children. They "touch" their world in the second half of the first year in life as do other children. In the second year of life they "understand" and from the third to the fourth years they really learn to "comprehend." It is absurd to provide aids or instructions to children afflicted with phocomelia before they have reached the proper age to understand and mentally digest the training measures. All training procedures should be in accordance with the child's age and should be adjusted to his degree of maturity.

j. Operative treatments should be postponed until the child has reached proper maturity. An extremity stump should never be amputated until it is absolutely certain that it is and will remain obstructive. It is possible that within a few years a slightly movable fingerbud at the shoulder can control a prosthesis which is operated by some external energy-source.

7. Shortly after birth, the children should be admitted to a special, properly equipped orthopedic clinic, erected as a center for the region, where medical consultations will take place. The general situation and future possibilities should be discussed with the parents prior to the child's admission to a neighboring children's clinic. A complete examination and a

thorough anamnestic research of the possible cause should be conducted at the clinic. It is also helpful when the parents can be shown another patient as an example of the development which can be expected of their child.

Treatment

Centralization of treatment centers into a few areas of the Federal Republic is desirable. The inquiry pertaining to a child's history should be limited to a few interrogators if dependable information about the possible causes of the malformation is to be secured. A large number of cases would be necessary for statistical significance. If useful results are to be obtained a very experienced staff is required to formulate remedial plans, exploit educational possibilities and arrange for the application of prosthetic and technical aids for phocomelia cases. Close contact between the attending and consulting physicians in the Federal Republic is essential in order to establish an effective defense against this catastrophe.

The children should be sent to kindergarten at an early age so that they will learn to move around and adjust to society. At the same time society should learn to associate with them and to respect them.

A constructive program for children with amputations and peromelia has been conducted and promoted in the United States for many years with large funds. We in the Federal Republic are just beginning to fill the gap which exists in the fabrication of prostheses for children. Almost all children's prostheses here are fabricated with adult components which are either too large or too heavy. Individual parts must of necessity be reduced or constructed according to the special requirements of children. Leg prostheses for children are relatively easy to fabricate. Artificial arms with grasping hooks and joints were prescribed in previous years only in puberty or to unilateral amputees who had become accustomed to their one-armed status and were rarely able to learn anew.

The experiences of the Americans has demonstrated that children should be provided with prostheses which can assist them to perform functions in accordance with their age group and degree of maturity. Children between one and one-and-a-half years of age with unilateral below-elbow peromelia are provided with simple aids so that they can crawl with arms of equal length. When the child wants to grasp at the age of three, he receives simple grasp instruments which he operates with the muscles of the shoulder girdle. More complicated prostheses are provided during the later stages which help to set higher standards for the child.

From a practical standpoint, this relatively simple program for unilateral and even for bilateral peromelia cases cannot be applied to the great number of newly-born children with bilateral severe phocomelia in the Federal Republic.

The 148 cases in our clinic presented in Table 2 are so severely damaged that 40 (almost 30%) have bilateral arm deformities from amelia to seal hands with short intercalary pieces which do not permit combined grasp-function with both extremity buds. This fact indicates that without aid they must depend on their feet, which are deformed in the same way in five cases.

These extremity buds, which are often grotesque, do not fit into any conventional prosthesis. Many could be utilized to control and make use of prostheses with grasp instruments by motion and sensation. Extended research and development studies are necessary, however, to construct these instruments and make them safe for use.

In many cases it is also an extensive task to guide the children to learn substitute performances with mouth or feet or trunk and shoulder motions.

This problem was previously handled only in specific cases in the large German institutions for the education of the disabled but we now must consider the problems of several hundred arm amputees who should not be allowed to waste away as nursing cases.

Conclusions

Let us suppose that all of the children with phocomelia with a long intercalary piece or with lesser degrees of bilateral arm deformities will be able to attend regular school but that 40 of the 148 cases with severe bilateral arm deformities in our small statistical survey will require care in institutional and special schools. This means that two or three groups of children will be ready to enter special schools in five or six years. The number of children is so great that our large institutions for the disabled are already making the necessary preparations.

Many of these children will require various kinds of technical aids during the coming years: arm prostheses which will be operated by their own physical strength or by external power from other energy resources, aids for eating, writing and body care, toys for exercises, leg prostheses and walking apparatus, self-propelled wheel chairs which will be developed for children with severe quadrilateral extremity deformities. These are tasks which the German orthopedic mechanics trade cannot handle properly as a sideline. The problems can only be mastered by a large-scale research and development program with training workshops.

A system of artificial arms has been developed and improved for a number of years at the Orthopedic University Clinic Schlierbach near Heidelberg. These arms are operated by compressed gas as an energy source—the Heidelberg pneumatic prosthesis. Our research workshop and school for the disabled has tried over a period of many years to fabricate artificial arms which would be useful for children. It is desirable that both clinics continue their close cooperation in a sustained effort to solve these acute problems.

In the majority of cases, children with phocomelia are given initial training in the use of their limb stumps. An attempt is then made to promote function with assistive apparatus. Later, a final apparatus or prosthesis which is individually practical can be fabricated.

Children grow up and require revisions in their prosthetic devices from time to time. It may be assumed that the second and later provisions can be handled by local orthopedic mechanics. The developers of special components and those who train orthopedic mechanics should work together closely in the construction of these aids.

As children mature, their requirements regarding technical aids for daily life and professional needs become more demanding. For many years to come, intensive work will be required from all professional groups concerned with the rehabilitation of disabled young people and, at the same time, institutions are greatly needed to assist the children to become accepted and productive members of society.