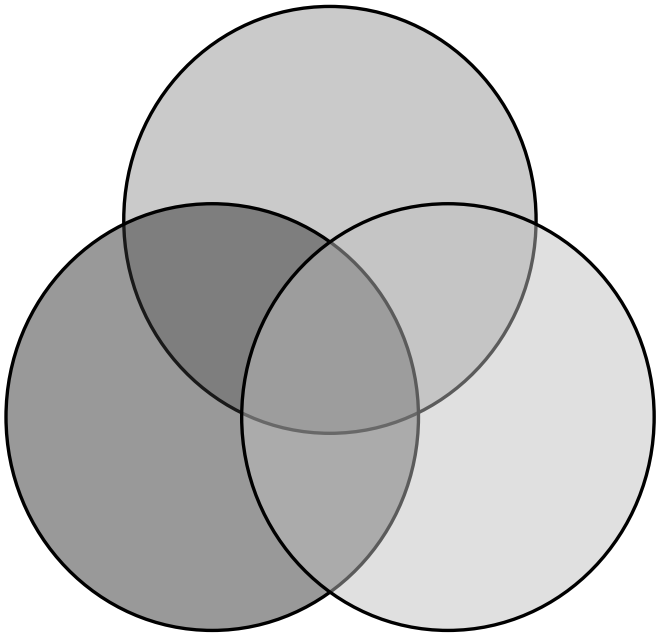
Diagnostic Guide for Fetal Alcohol Spectrum Disorders

The 4-Digit Diagnostic Code

Third Edition
2004



FAS Diagnostic and Prevention Network
University of Washington
Seattle Washington

Diagnostic Guide for Fetal Alcohol Spectrum Disorders: The 4-Digit Diagnostic Code

Third Edition

2004.

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Preface

What's New in this Third Edition?

The first and second editions of the Diagnostic Guide were printed in 1997 and 1999 (Astley and Clarren, 1997, 1999). The key updates in this third edition are presented below. These updates are based on our use of the 4-Digit Code for the past seven years on over 2,000 patients, advancements in medical research, U.S. and Canadian efforts to establish National Diagnostic Guidelines, and feedback from over 70 clinical teams trained to use the 4-Digit Diagnostic Code. We will continue to make modifications that enhance accuracy, improve clarity, and increase ease of use. We hope you will find this comprehensive approach to the diagnosis of individuals with prenatal alcohol exposure helpful and broadly applicable.

Key updates in this 3rd edition include:

1. Re-Classification of Nineteen 4-Digit Codes across Seven Diagnostic Categories. Based on current efforts in the U.S. and Canada to establish National Diagnostic Guidelines, and our own experience using the 4-Digit Code, we have reclassified 19 of the 246 4-Digit Codes. Most of these reclassifications reflect the widespread consensus to relax the growth criteria. A detailed presentation of which codes were reclassified, why they were reclassified, and the impact the reclassification has on the prevalence of each diagnostic category can be found on the FAS DPN website (http://depts.washington.edu/fasdpn).

2. Modification of the growth deficiency case-definitions to harmonize with the U.S. and Canadian Diagnostic case-definitions for growth deficiency. This modification allows one to document and differentiate growth deficiency at both the 3rd and 10th percentiles.

3. Updated FASD Diagnostic Form with a new Functional Domains page. The FASD Diagnostic Form has been updated to provide a more comprehensive format. An additional page has been added to allow one to document "Domains of Brain Dysfunction". Documentation of impaired domains (e.g., cognition, memory, executive function, etc.) is a key component of the Canadian and U.S. National Diagnostic Guidelines and has always been required to derive/support a CNS Rank 3 classification when using the 4-Digit Code.

4. Updated Growth Charts. The most recent 2000 CDC growth charts are included with reference to their website for computerized charting of growth.

5. New Caucasian and African American Lip-Philtrum Guides, 2004. A new Caucasian Lip-Philtrum Guide was printed that uses higher-resolution, higher quality photographs. The magnitude of lip thinness and philtrum smoothness remain unchanged from the 1999 Caucasian Lip-Philtrum Guide. A new African American Lip-Philtrum Guide has also been created. The cut-off values for each of the five ranks in the African American Guide were set to be comparable to the percentile cutoffs used in the Caucasian Lip-Philtrum Guide. Both Guides require a Rank 4 or 5 lip and philtrum to meet the criteria for the FAS facial phenotype. The 2004 modified growth table is printed on the backside of each Lip-Philtrum Guide.

I. Introduction

A. What are Fetal Alcohol Syndrome (FAS) and Fetal Alcohol Spectrum Disorders (FASD)

FAS is a permanent birth defect syndrome caused by maternal consumption of alcohol during pregnancy. The definition of the FAS has changed little since the 1970's when the condition was first described and refined (Jones and Smith, 1973; Rosett, 1980; Clarren and Smith, 1978; Sokol and Clarren, 1989; Stratton *et al*., 1996). The condition has been broadly characterized by prenatal and/or postnatal growth deficiency, a unique cluster of minor facial anomalies, and central nervous system (CNS) abnormalities. FAS is the leading known cause of mental retardation/developmental disabilities in the Western World (Abel & Sokol, 1987) and is entirely preventable. The prevalence of FAS is estimated to be 1 to 3 per 1,000 live births (Stratton et al., 1996) in the general population, but has been documented to be as high as 10 to 15 per 1,000 in some high-risk populations (Astley et al., 2002).

The physical, cognitive, and behavioral deficits observed among individuals with prenatal alcohol exposure are not dichotomous, that is either normal or clearly abnormal. Rather, the outcomes, and the prenatal alcohol exposure, all range along separate continua from normal to clearly abnormal and distinctive. This full range of outcomes observed among individuals with prenatal alcohol exposure has come to be called Fetal Alcohol Spectrum Disorders (FASD). The term FASD is not intended for use as a clinical diagnosis. A patient would not receive a diagnosis of FASD, for the term is too broadly defined to be of clinical value. FAS, on the other hand, is a clinical diagnosis and is one of several alcohol-related diagnoses that fall under the umbrella of FASD.

Although reference to the harmful effects of prenatal alcohol exposure on infant outcome dates back to the biblical literature, it was not until 1968 when the first reference was published in the medical literature by Lemoine and colleagues from France (Lemoine et al., 1968). Ulleland and colleagues from the United States published similar research findings in 1970 and 1972 (Ulleland et al., 1970; Ulleland, 1972). Using today's terminology, one could say Lemoine and Ulleland were the first to describe FASD in the medical literature. In 1973, Jones and Smith coined the term FAS (Jones & Smith, 1973) to describe a subset of alcohol-exposed children, obtained from Dr. Ulleland's study and their own clinical records, who shared a common pattern of malformation (Jones et al., 1973).

B. The Diagnostic Challenge

FASD can present a daunting, but not insurmountable challenge for diagnosis. Individuals with prenatal alcohol exposure present with a wide range of outcomes, most of which are not specific to prenatal alcohol exposure and often manifest differently across the lifespan. Professionals from multiple disciplines (medicine, psychology, speech-language pathology, occupational therapy, etc.) are needed to accurately assess and interpret the broad array of outcomes that define the diagnoses. The pattern and severity of outcome is dependent on the timing, frequency, and quantity of alcohol exposure (which is rarely known with any level of accuracy), and is frequently confounded by other adverse prenatal and postnatal exposures and events.

In the absence of accurate, precise, and unbiased methods for measuring and recording the severity of exposures and outcomes in individual patients, diagnoses have varied widely from clinic to clinic

(Aase, 1994; Astley & Clarren 2000; Chavez et al., 1988; Stratton et al., 1996). From a clinical perspective, diagnostic misclassification leads to inappropriate patient care, increased risk for secondary disabilities (Streissguth & Kanton, 1997) and missed opportunities for primary prevention. From a public health perspective, diagnostic misclassification leads to inaccurate estimates of incidence and prevalence (Stratton et al., 1996). Inaccurate estimates thwart efforts to allocate sufficient social, educational, and health care services to this high-risk population, and preclude accurate assessment of primary prevention intervention efforts. From a clinical research perspective, diagnostic misclassification reduces the power to identify clinically meaningful contrasts between FAS and control groups (Astley & Clarren, 2001). Non-standardized diagnostic methods prevent valid comparisons between studies.

The 4-Digit Diagnostic Code was originally created in 1997 to address the following limitations in the conventional gestalt approach to diagnosing individuals with prenatal alcohol exposure.

1. *There have been no standardized operational definitions for FAS or for any of the other diagnoses that fall under the umbrella of FASD. Rather, there have been diagnostic guidelines that physicians have been encouraged to follow, but the guidelines have not been sufficiently specific to assure diagnostic accuracy or precision.*

   For example, according to the diagnostic guidelines published by Sokol and Clarren (1989), which were a minor modification of the 1980 definition of FAS by the Fetal Alcohol Study Group of the Research Society for Alcoholism (Rosett, 1980), which, in turn, were derived from the work of Clarren and Smith (1978): "The diagnosis of FAS can only be made when the patient has signs of abnormality in each of the three categories: 1) Prenatal and/or postnatal growth retardation [weight and/or length below the 10^{th} percentile when corrected for gestational age], 2) central nervous system involvement (including neurological abnormality, developmental delay, behavioral dysfunction or deficit, intellectual impairment, and/or structural abnormalities, such as microcephaly [head circumference below the 3^{rd} percentile or brain malformations found on imaging studies or autopsy] and 3) a characteristic face, currently qualitatively described as including short palpebral fissures, an elongated midface, a long and flattened philtrum, thin upper lip, and flattened maxilla."

   The 1996 guidelines for the diagnosis of FAS proposed by the Institute of Medicine (Stratton et al., 1996) took a similar approach. The diagnosis of FAS can be made when the patient presents with: "1) Evidence of growth retardation, as in at least one of the following: a) low birth weight for gestational age; b) decelerating weight over time not due to nutrition; or c) disproportional low weight to height; 2) Evidence of a characteristic pattern of facial anomalies that includes features such as short palpebral fissures and abnormalities in the premaxillary zone (e.g., flat upper lip, flattened philtrum, and flat midface); and 3) Evidence of CNS neurodevelopmental abnormalities, as in at least one of the following: a) decreased cranial size at birth; b) structural brain abnormalities (e.g., microcephaly, partial or complete agenesis of the corpus callosum, cerebellar hypoplasia);c) neurological hard or soft signs (as age appropriate), such as impaired fine motor skills, neurosensory hearing loss, poor tandem gait, poor eye-hand coordination."

   Although these descriptions do provide guidance, they are not sufficiently specific to assure diagnostic accuracy and precision. They reflect a more "gestalt" approach to diagnosis. The guidelines for CNS abnormalities do not address how many areas of deficit must be present, how

severe the deficits must be, or what level of documentation must exist to substantiate the presence of the deficit. The guidelines for the facial phenotype are equally nonspecific. How many facial features must be present, how severe must the features be, and what scale of measurement should be used to judge the severity? One need only read the clinical literature or review medical records, birth certificates, birth defect registries or ICD-9 codes to see how variably these criteria are interpreted, applied and reported (CDC, 1995, 1995a; Cordero et al., 1994; Ernhart et al., 1995; Stratton et al., 1996).

New U. S. diagnostic guidelines for FAS (Bertrand et al., 2004) and Canadian diagnostic guidelines for FASD (Chudley et al., 2004) offer more standardized, case-defined criteria than those published in previous guidelines (Sokol and Clarren, 1989, Stratton et al., 1996). Both are slated for release in 2004.

2. *There has been a lack of objective, quantitative scales to measure and report the magnitude of expression of key diagnostic features*

   For example, although a thin upper lip and smooth philtrum are key diagnostic features (Astley & Clarren, 1996; Clarren & Smith, 1978; Jones & Smith, 1973; Smith, 1979; Stratton et al., 1996), quantitative measurement scales were never used to measure thinness or smoothness, and guidelines had never been established for how thin or smooth the features must be. Objective quantitative scales not only improve accuracy and precision, but also establish a common numeric language for communicating outcomes in medical records and in the medical literature.

3. *The term fetal alcohol effects (FAE) was broadly used and poorly defined.*

   The term 'suspected fetal alcohol effects' was first introduced into the medical literature in 1978 and was defined as 'less complete partial expressions' of FAS in individuals with prenatal alcohol exposure (Clarren & Smith, 1978). Based on this definition, an individual whose mother drank a few glasses of wine intermittently throughout pregnancy and presented with attention deficit hyperactivity disorder would meet the criteria for FAE. So would an individual whose mother drank a fifth of vodka daily throughout pregnancy and presented with microcephaly, severe mental retardation, growth deficiency and no facial anomalies. The broad use of this term and the reluctance to abandon it points to the clear need to develop diagnostic terms for individuals with prenatal alcohol exposure who present with physical anomalies and/or cognitive/behavioral disabilities, but do not meet the criteria for FAS. New diagnostic terms that more finely differentiate the variable exposures and outcomes of individual patients, without implying alcohol as the sole causal agent, are needed.

4. *Clinical terms like FAE* (Aase et al., 1995)*, alcohol-related birth defects (ARBD) (Stratton et al., 1996) and alcohol-related neurodevelopmental disorder (ARND) (Stratton et al., 1996) imply a causal link between alcohol exposure and outcome in a given individual that, to date, cannot be medically confirmed. Leading dysmorphologists in the field of FAS diagnosis have formally requested that the term FAE no longer be used for this reason (Aase et al., 1995; Sokol & Clarren, 1989).*

   With the likely exception of the full facial phenotype, no other physical anomalies or cognitive/behavioral disabilities observed in an individual with prenatal alcohol exposure are necessarily specific to (caused only by) their prenatal alcohol exposure (Stratton et al., 1996). Features

such as microcephaly, neurological abnormalities, attention deficit, mental retardation, and growth deficiency frequently occur in individuals with prenatal alcohol exposure, and frequently occur in individuals with no prenatal alcohol exposure. The diagnostic terms ARBD and ARND introduce the same limitation as does FAE, namely, implying alcohol exposure caused the birth defect or neurobehavioral disorder in an individual patient. The 4-Digit Code avoids this problem by using a nomenclature that reports the patient was *exposed* to prenatal alcohol rather than reporting the patient's outcomes are *alcohol effects* or *alcohol-related outcomes*. The 4-Digit Code also requires that all other adverse prenatal and postnatal exposures and events be documented for they too serve as important risk factors that must be taken into consideration when deriving a diagnosis and intervention plan.

5. *Too often diagnoses depicting FASD are reported in the medical records and medical literature with no documentation of the method used to derive the diagnosis and little or no documentation of the data used to support the diagnosis.*

   Failure to report this information can limit the patient's ability to qualify for and receive appropriate intervention services from subsequent health care, social service, and educational providers. For example, simply reporting that an individual has FAS does little to convey the individual's strengths and disabilities. Some individuals with FAS have low IQs, some have normal IQs, some have attention deficits, some do not, some have problems with memory, while others have language deficits. From a public health perspective, failure to report these data also prevents surveillance efforts from accurately tracking the prevalence of FASD diagnoses in the population. The supportive data are needed to validate the diagnoses. Accurate surveillance is vital for setting public health policy and assessing the effectiveness of primary prevention efforts. The 4-Digit Code requires that data be collected not just to support the diagnosis, but to derive the diagnosis. The 4-Digit Code provides a comprehensive FASD Diagnostic Form for recording all supportive data and provides a numeric classification scheme that is readily incorporated into clinical, research, and surveillance databases.

C. Meeting the Diagnostic Challenge

Each of the above limitations has been largely overcome with the development of the "*4-Digit Diagnostic Code*". The four digits reflect the magnitude of expression of four key diagnostic features of FASD in the following order: (1) growth deficiency, (2) the FAS facial phenotype, (3) CNS abnormalities, and (4) prenatal alcohol exposure. The magnitude of expression of each feature is ranked independently on a 4-point Likert scale with 1 reflecting complete absence of the FAS feature and 4 reflecting a strong "classic" presence of the FAS feature. Thus, the 4-Digit Code 4444 reflects the most severe expression of FAS (significant growth deficiency, all three FAS facial features, structural/neurological evidence of CNS damage, and confirmed prenatal exposure to high levels of alcohol). At the opposite end of the scale is the 4-Digit Code 1111 reflecting normal growth, none of the three FAS facial features, no evidence of CNS abnormalities, and confirmed absence of prenatal alcohol exposure. Every combination of 4-Digit Code has been observed in the Washington State FAS Diagnostic & Prevention Network.

This diagnostic method was developed through the combined expertise of the University of Washington FAS Diagnostic and Prevention Network (FAS DPN) interdisciplinary clinical team

(Clarren & Astley, 1997; Clarren et al., 2000) and the comprehensive records of over 2,000 patients (birth to 53 years of age) diagnosed through the FAS DPN.

D. Benefits of the 4-Digit Diagnostic Code

The 4-Digit Diagnostic Code:

1. Greatly increases diagnostic precision and accuracy through the use of objective, quantitative measurement scales, image analysis software, and specific case definitions.
2. Diagnoses the full spectrum of outcomes (FASD) observed in individuals of all ages with prenatal alcohol exposure.
3. Offers an intuitively logical numeric approach to reporting outcomes and exposure that reflects the true diversity and continuum of disability associated with prenatal alcohol exposure.
4. Documents the presence of prenatal alcohol exposure without judging its causal role.
5. Documents all other prenatal and postnatal adverse exposures and events that can also impact outcome.
6. Provides a quantitative measurement and reporting system that can be used independent of diagnostic nomenclature.
7. Can be taught to a wide array of health care and social service providers, thus greatly expanding the availability of diagnostic services. (Appendix 1)

The 4-Digit Code currently serves as the cornerstone of a fully integrated and highly successful screening, diagnostic, prevention and surveillance program in Washington State (Astley et al., 2002; Astley, 2004).

While this document might at first appear overly complex and perhaps daunting, one will find that this diagnostic approach is logical and easy to use, and will greatly facilitate the proper description and classification of patients presenting with all possible combinations of outcomes and exposures.

E. Other Syndromes

The methods of diagnosing fetal alcohol syndrome arise from the larger fields of teratology and dysmorphology (clinical genetics). It is essential to remember that many birth defect syndromes share *isolated* features, but each is differentiated by a unique *constellation* of features. A few examples of conditions that share some, but not all, of the features of FAS include fetal hydantoin syndrome, maternal PKU fetal effects, and fetal valproate syndrome. Although this guide is "FASD-specific", this in no way should imply that the diagnostician need not consider alternate or co-existing syndromic, medical or psychiatric conditions at all times. A differential diagnosis is essential in making an accurate diagnosis.

III. Instructions for Deriving the 4-Digit Code

A. The 4-Digit Diagnostic Code

What are the 4 Digits?

The four digits reflect the magnitude of expression of the four key diagnostic features of FASD in the following order: (1) growth deficiency, (2) the FAS facial phenotype, (3) CNS abnormalities, and (4) prenatal alcohol exposure. The 4-Digit Diagnostic Code is generated at the completion of the diagnostic evaluation using information recorded on the FASD Diagnostic Form. The code is derived following the directions in Sections III. B. 1 through B. 4.

4-Digit Diagnostic Code Grid

				3	4	4		4		
Severe	Severe	Definite	(4)		X	X		X	(4)	High risk
Moderate	Moderate	Probable	(3)	X					(3)	Some risk
Mild	Mild	Possible	(2)						(2)	Unknown
None	None	Unlikely	(1)						(1)	No Risk
Growth Deficiency	**FAS Facial Features**	**CNS Damage**		Growth	Face	CNS		Alcohol		**Prenatal Alcohol**

The 4-Digit Diagnostic Code 3444 inserted in the grid is one of twelve 4-Digit Codes that meet the diagnostic criteria for FAS.

How are the 4 Digits Ranked?

The magnitude of expression of each feature is ranked independently on a 4-point Likert scale with 1 reflecting complete absence of the FAS feature and 4 reflecting a strong "classic" presence of the FAS feature. Specific guidelines for ranking the magnitude of each of the FAS features are presented in Section III.B.

How Many 4-Digit Diagnostic Codes are There?

There are 256 possible 4-Digit Diagnostic Codes ranging from 1111 to 4444. The 256 codes and their corresponding clinical names are listed in numerical order in Section VI.

How Many Different Clinical Diagnostic Categories are There?

Each 4-Digit Diagnostic Code falls into one of 22 unique Clinical Diagnostic Categories (labeled A through V). A list of the 22 Diagnostic Categories is presented in Section IV. A list of the 4-Digit Diagnostic Codes, which fall within each Clinical Diagnostic Category, is presented in Section V.

What are the Names of the Clinical Diagnostic Categories?

The following terms are used in varying combinations to name the 22 diagnostic categories. They include:

- **Sentinel Physical findings:**
  The term "S*entinel Physical Findings*" is used in this diagnostic system when the patient presents with growth deficiency at the Rank 3 or 4 level and/or presents with the FAS facial phenotype at the Rank 3 or 4 level. The adjective *"sentinel"* refers to physical findings that are key diagnostic features of FAS. These include a unique cluster of minor facial anomalies (short palpebral fissures, thin upper lip, and a smooth philtrum) and growth deficiency. Other physical findings (major or minor anomalies) may be detected instead of or in addition to these sentinel findings that may suggest alternate or additional conditions. There are places on the Diagnostic Form to record and interpret other physical findings.

- **Static Encephalopathy:**
  The term *"encephalopathy"* refers to "any significant abnormal condition of the structure or function of brain tissues" (Anderson, 2002). The term *"static"* means that the abnormality in the brain is unchanging; neither progressing nor regressing. The term *"Static Encephalopathy"* is used in this diagnostic system when the patient presents with significant structural, neurological, and/or functional abnormalities that strongly support the presence of underlying CNS damage at the Rank 3 and/or Rank 4 levels. The term does not define or suggest any specific pattern of structural, neurological, or functional abnormality.

- **Neurobehavioral Disorder:**
  The term *"Neurobehavioral Disorder"* is used in this diagnostic system when the patient presents with cognitive/behavioral dysfunction at the Rank 2 level and no evidence of structural, neurological or functional abnormalities at the Rank 3 or Rank 4 levels.

- **Alcohol (Exposed, Not Exposed, Exposure Unknown):**
  These terms are used to reflect prenatal alcohol exposure and its potential risk to the unborn child. Alcohol exposure is reported <u>independently</u> of outcome(s) and does not imply that a causal association exists between the exposure and the outcome(s).

- **Fetal Alcohol Syndrome (alcohol exposed)**
  The term FAS is used to refer to patients who present with one of twelve 4-Digit Diagnostic Code combinations reflecting growth deficiency; the full FAS facial phenotype; significant structural, neurological, and/or functional CNS abnormalities; and confirmed prenatal alcohol exposure. These 12 Codes are presented in Section V.

- **Fetal Alcohol Syndrome (alcohol exposure unknown)**
  A diagnosis of FAS can be rendered when prenatal alcohol exposure is "unknown" but only when the outcomes (growth, face, and CNS) are at the severe end of the spectrum to maintain the specificity of these outcomes to prenatal alcohol exposure. (Astley et al., 2001) Six 4-Digit Codes fall under this category (Section V).

- **Partial Fetal Alcohol Syndrome (alcohol exposed):**
  This term is used for patients who present with static encephalopathy, most (but not all) of the growth and/or facial features of FAS, and have a confirmed history of prenatal alcohol exposure. Given the fact that variable presentation is the rule rather than the exception after teratogenic exposures, we felt it was appropriate to establish this diagnostic category. Twenty 4-Digit Codes fall under this category (Section V).

- **Fetal Alcohol Syndrome Phenocopy (no alcohol exposure):**
  This term is used for patients who meet the growth, face and CNS criteria for FAS, but have a confirmed absence of alcohol exposure during gestation. We have never seen such a case (or phenocopy), but we may some day.

The names assigned to each diagnostic category reflect the patient's clinical outcome and alcohol exposure. The names are listed in Sections IV and V. The first three categories (A through C) meet the criteria for a clinical diagnosis of FAS and are named as such. The fourth category (D) applies to the patient who presents with all of the features of FAS, but has a confirmed *absence* of prenatal alcohol exposure from conception to birth. This category is referred to as a FAS Phenocopy and has yet to be observed. The remaining 19 categories (E through V) do not meet the minimum criteria for FAS or partial FAS. These are subsequently named to reflect the Likert ranking of each digit in the 4-Digit Diagnostic Code. For example, a code of 3243 is the Diagnostic Category called "*Sentinel physical finding(s) / static encephalopathy (alcohol exposed)*".

Which Diagnostic Categories are Comparable to PFAE, ARND and ARBD?

Many 4-Digit Codes within Diagnostic Categories E through I would previously have been referred to as "possible fetal alcohol effects" (PFAE), "alcohol-related birth defects" (ARND) or "alcohol-related neurodevelopmental disorder" (ARBD). (Sokol & Clarren, 1989; Stratton et al., 1996) A report that translates which 4-Digit Codes meet the criteria for ARND and ARBD can be found on the FAS DPN website http://depts.washington.edu/fasdpn. Categories J through V are categories that describe a large number of patient groups who have never been adequately classified or described by previous FASD diagnostic guidelines.

Ultimately, establishing terms that are both clinically accurate, broadly applicable, and facilitate access to services remains a challenge. It is important to remember that the 4-Digit Code provides a numeric measurement and reporting system for exposures and outcomes that can be used independently of the proposed diagnostic nomenclature.

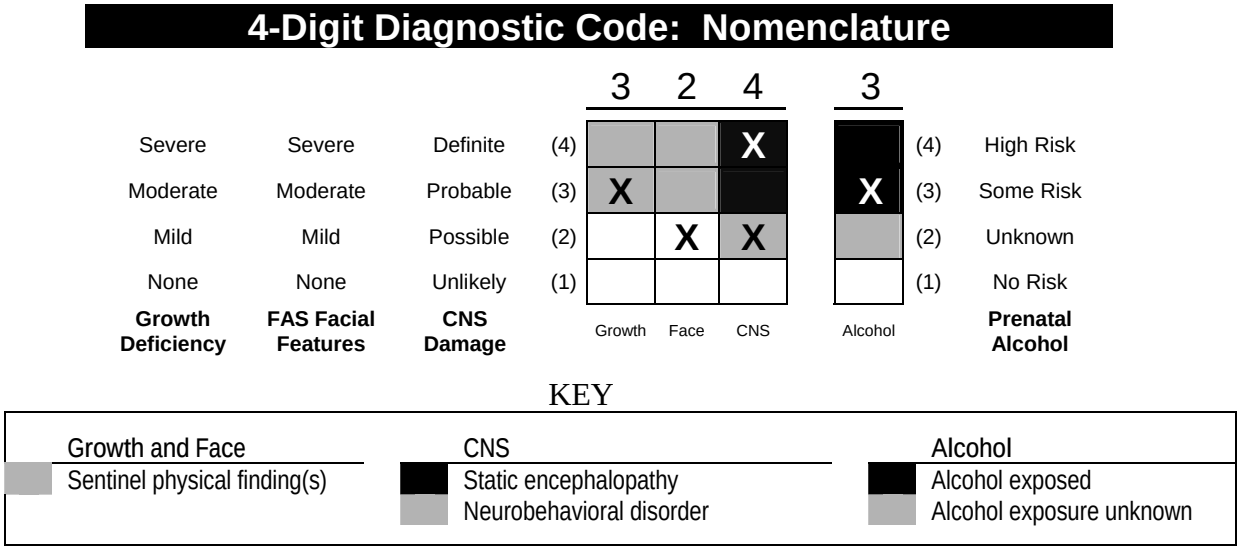
How are the Names of the Clinical Diagnostic Category Constructed?

- Growth deficiency and facial features are physical features. When either feature receives a rank of 3 or 4, *Sentinel physical finding(s)* is placed at the beginning of the name.

- When CNS receives only a Rank 2, the term *Neurobehavioral Disorder* is included in the name. When CNS receives a Rank 3 or 4, the term *Static Encephalopathy* is included in the name.

- When alcohol exposure receives a Rank 3 or 4, (*alcohol exposed*) is placed at the end of the name. When alcohol exposure receives a Rank 2, (*alcohol exposure unknown*) is placed at the end of the name.

- When the criteria for FAS or PFAS are met, those clinical terms are used in place of the more generic terms. For example the term FAS is used rather than *Sentinel physical finding(s / static encephalopathy (alcohol exposed).*



The 4-Digit Code 3243 would receive the clinical name *Sentinel physical finding(s) / static encephalopathy (alcohol exposed).* Note that the CNS received both Rank 4 and Rank 2. The higher Rank is used to derive the 4-Digit Code and construct the name. A code of 1222 would receive the clinical name *Neurobehavioral disorder (alcohol exposure unknown).*

How Do You Explain the Diagnosis to the Patient?

Generic summaries of each of the 22 Clinical Diagnostic Categories are presented in Section VII. These summaries can be used as the first page of the patient's final Medical Summary Note. Subsequent pages in the Medical Summary Note should document the findings and recommendations specific to the patient. We recommend the growth, face, CNS, and exposure data, used to generate the 4-Digit Code, be reported in the Medical Summary Note to provide essential information to subsequent medical professionals and facilitate records-based public health surveillance efforts.

IV. Diagnostic Categories

The 256 Diagnostic Codes can be logically grouped into 22 Diagnostic Categories

Category	Name
A	Fetal alcohol syndrome (alcohol exposed)
B	Fetal alcohol syndrome (alcohol exposure unknown)
C	Partial fetal alcohol syndrome (alcohol exposed)
D	Fetal alcohol syndrome phenocopy (no alcohol exposure)
E	Sentinel physical finding(s) / static encephalopathy (alcohol exposed)
F	Static encephalopathy (alcohol exposed)
G	Sentinel physical finding(s) / neurobehavioral disorder (alcohol exposed)
H	Neurobehavioral disorder (alcohol exposed)
I	Sentinel physical finding(s) (alcohol exposed)
J	No sentinel physical findings or CNS abnormalities detected (alcohol exposed)
K	Sentinel physical finding(s) / static encephalopathy (alcohol exposure unknown)
L	Static encephalopathy (alcohol exposure unknown)
M	Sentinel physical finding(s) / neurobehavioral disorder (alcohol exposure unknown)
N	Neurobehavioral disorder (alcohol exposure unknown)
O	Sentinel physical finding(s) (alcohol exposure unknown)
P	No sentinel physical findings or CNS abnormalities detected (alcohol exposure unknown)
Q	Sentinel physical finding(s) / static encephalopathy (no alcohol exposure)
R	Static encephalopathy (no alcohol exposure)
S	Sentinel physical finding(s) / neurobehavioral disorder (no alcohol exposure)
T	Neurobehavioral disorder (no alcohol exposure)
U	Sentinel physical finding(s) (no alcohol exposure)
V	No sentinel physical findings or CNS abnormalities detected (no alcohol exposure)

V. 4-Digit Diagnostic Codes
Within each Diagnostic Category

Category	Diagnostic Name and Codes
A	Fetal alcohol syndrome (alcohol exposed) 2433 3433 4433 2434 3434 4434 2443 3443 4443 2444 3444 4444
B	Fetal alcohol syndrome (alcohol exposure unknown) 2432 3432 4432 2442 3442 4442
C	Partial fetal alcohol syndrome (alcohol exposed) 1333 1433 2333 3333 4333 1334 1434 2334 3334 4334 1343 1443 2343 3343 4343 1344 1444 2344 3344 4344
D	Fetal alcohol syndrome phenocopy (no alcohol exposure) 3431 4431 3441 4441
E	Sentinel physical finding(s) / static encephalopathy (alcohol exposed) 3133 3233 4133 4233 3134 3234 4134 4234 3143 3243 4143 4243 3144 3244 4144 4244
F	Static encephalopathy (alcohol exposed) 1133 1233 2133 2233 1134 1234 2134 2234 1143 1243 2143 2243 1144 1244 2144 2244
G	Sentinel physical finding(s) / neurobehavioral disorder (alcohol exposed) 1323 2323 3123 3323 4123 4323 1324 2324 3124 3324 4124 4324 1423 2423 3223 3423 4223 4423 1424 2424 3224 3424 4224 4424

VI. 4-Digit Diagnostic Codes
Sorted Numerically

Code	Category	Diagnostic Name
1111	V	No sentinel physical findings or CNS abnormalities detected (no alcohol exposure)
1112	P	No sentinel physical findings or CNS abnormalities detected (alcohol exposure unk.)
1113	J	No sentinel physical findings or CNS abnormalities detected (alcohol exposed)
1114	J	No sentinel physical findings or CNS abnormalities detected (alcohol exposed)
1121	T	Neurobehavioral disorder (no alcohol exposure)
1122	N	Neurobehavioral disorder (alcohol exposure unknown)
1123	H	Neurobehavioral disorder (alcohol exposed)
1124	H	Neurobehavioral disorder (alcohol exposed)
1131	R	Static encephalopathy (no alcohol exposure)
1132	L	Static encephalopathy (alcohol exposure unknown)
1133	F	Static encephalopathy (alcohol exposed)
1134	F	Static encephalopathy (alcohol exposed)
1141	R	Static encephalopathy (no alcohol exposure)
1142	L	Static encephalopathy (alcohol exposure unknown)
1143	F	Static encephalopathy (alcohol exposed)
1144	F	Static encephalopathy (alcohol exposed)
1211	V	No sentinel physical findings or CNS abnormalities detected (no alcohol exposure)
1212	P	No sentinel physical findings or CNS abnormalities detected (alcohol exposure unk.)
1213	J	No sentinel physical findings or CNS abnormalities detected (alcohol exposed)
1214	J	No sentinel physical findings or CNS abnormalities detected (alcohol exposed)
1221	T	Neurobehavioral disorder (no alcohol exposure)
1222	N	Neurobehavioral disorder (alcohol exposure unknown)
1223	H	Neurobehavioral disorder (alcohol exposed)
1224	H	Neurobehavioral disorder (alcohol exposed)
1231	R	Static encephalopathy (no alcohol exposure)
1232	L	Static encephalopathy (alcohol exposure unknown)
1233	F	Static encephalopathy (alcohol exposed)
1234	F	Static encephalopathy (alcohol exposed)
1241	R	Static encephalopathy (no alcohol exposure)
1242	L	Static encephalopathy (alcohol exposure unknown)
1243	F	Static encephalopathy (alcohol exposed)
1244	F	Static encephalopathy (alcohol exposed)
1311	U	Sentinel physical finding(s) (no alcohol exposure)
1312	O	Sentinel physical finding(s) (alcohol exposure unknown)
1313	I	Sentinel physical finding(s) (alcohol exposed)
1314	I	Sentinel physical finding(s) (alcohol exposed)
1321	S	Sentinel physical finding(s) / neurobehavioral disorder (no alcohol exposure)
1322	M	Sentinel physical finding(s) / neurobehavioral disorder (alcohol exposure unknown)
1323	G	Sentinel physical finding(s) / neurobehavioral disorder (alcohol exposed)